
New York State Department of Health
Birth Defects Registry

ICD-10 Coding Manual

List of all Reportable Congenital Malformations

Last updated 10/18/2021

Table of Contents

Reporting Requirements and Instructions	- 3 -
Children to Report:.....	- 3 -
What to Report:	- 3 -
Common Acronyms:.....	- 3 -
Color Coding:.....	- 3 -
Common Notation:.....	- 3 -
Congenital Malformations of the Nervous System (Q00-Q07)	- 4 -
Congenital Malformations of Eye, Ear, Face and Neck (Q10-Q18).....	- 11 -
Congenital Malformations of the Circulatory System (Q20-Q28).....	- 17 -
Congenital Malformations of the Respiratory System (Q30-Q34)	- 24 -
Congenital Malformations of the Cleft Lip and Cleft Palate (Q35-Q37)	- 27 -
Congenital Malformations of the Digestive System (Q38-Q45)	- 29 -
Congenital Malformations of Genital Organs (Q50-Q56)	- 35 -
Congenital Malformations of the Urinary System (Q60-Q64).....	- 40 -
Congenital Malformations of the Musculoskeletal System (Q65-Q79)	- 43 -
Other Congenital Malformations (Q80-Q89)	- 60 -
Chromosomal Abnormalities, Not Elsewhere Classified (NEC) (Q90-Q99).....	- 66 -
Other Selected Reportable Conditions.....	- 70 -
Stillbirth Codes (O36.4, Z37)	- 76 -
Revision History	- 77 -

Reporting Requirements and Instructions

Children to Report:

Hospitals and healthcare providers are required to report children born or living in New York State (NYS) who have been diagnosed, before two years of age (unless otherwise specified), with birth defects, chromosomal anomalies, or were stillborn or other non-liveborn. Certain birth defects are required to be reported for children up to age 10 (specified in red). For purposes of this registry, a birth defect (also referred to as anomaly or malformation) is defined as any structural, functional, and/or specific biochemical abnormalities which were determined genetically or induced during gestation and not due to birthing events. This includes newborns and pediatric patients with a birth defect, even if they are transferred to or from another hospital, and any child with a birth defect who dies in the hospital (inpatient or emergency room).

What to Report:

This coding manual contains a complete list of NYS Birth Defects Registry (BDR) reportable anomalies. Rows with grey backgrounds represent non-billable codes identified by the ICD-10 nomenclature. When reporting syndromes which have several defects, report each individual defect as well as the associated syndrome. Tables contain the following three columns:

1st Column called ‘ICD-10’ – ICD-10 codes that correspond to the child’s birth defect.

2nd Column called ‘Description’ – Lists the standard narrative birth defect description(s) and may provide specific directions (e.g., must specify, female only, male only, birth weight criteria, etc.)

- All major defects and stillbirths must be reported
 - Minor defects, descriptions with an *(m)* (e.g. *(m) ankyloglossia*), are only required to be reported if the child also has one or more major defects or was stillborn or other non-liveborn.
- Some birth defects are only reportable if the child is a certain sex or was born above a certain birth weight. Please only report them if they meet the given criteria or the criteria is unknown. Examples:
 - Only report patent ductus arteriosus (PDA) if the child’s birth weight is 1,500 grams or greater. PDA is normal and necessary as a fetus develops and should close at birth.
 - Only report K40-, inguinal hernia, for female children
- **Please include as much specific detail from the medical record as possible for each defect**
 - Indicate the laterality if applicable (e.g., talipes equinovarus, left)
 - Avoid reporting vague descriptions starting with “Other” or those with codes ending in -8 or -9
 - Add more specificity if you see ‘**(must specify)**’ next to an ICD-10 Description

NOTE: Specific narrative descriptions of birth defects are required for reporting of all birth defects.

3rd Column called ‘ICD-9’: ICD-9 code that corresponds with the respective ICD-10 code.

Syndrome and chromosomal defects: please report the cytogenetic diagnoses or test results. These results are also required for defects under ICD-10 code Q56 (indeterminate sex or pseudohermaphroditism).

Common Acronyms:

(m) = Minor defect

NOS = Not Otherwise Specified

NEC = Not Elsewhere Classified

Color Coding:

Black font: From ICD-10 Codebook

Blue font: Additional synonyms or descriptions of other conditions also included under this code.

Red font: Key instructions to follow

Common Notation:

Includes: Further defines, or gives examples of, the content of the code or category

Excludes 1 (Type 1 Excludes): Indicates the code identified under the ‘excludes’ statement and the respective ICD10 code cannot be reported together because the two conditions cannot appear together.

Excludes 2 (Type 2 Excludes): Condition excluded is not part of the condition it is excluded from, but a patient may have both conditions at the same time. Report both codes if the patient has both conditions.

Please see [Revision History](#) section for changes in ICD-10 codes over time.

Congenital Malformations of the Nervous System (Q00-Q07)

ICD-10	Description	ICD-9
Q00	Anencephaly and similar malformations	
Q00.0	Anencephaly/Anencephalus Acephaly Acrania Amyelencephaly/Amyelencephalus Congenital brain aplasia Hemianencephaly Hemicephaly	740.0
Q00.1	Craniorachischisis (fissure skull/vertebral column)	740.1
Q00.2	Iniencephaly	740.2
Q01	Encephalocele Includes: Arnold-Chiari syndrome, type III, encephalocystocele, encephalomyocele, hydroencephalocele, hydromeningocele (cranial), meningocele (cerebral), meningoencephalocele Excludes 1: Meckel-Gruber syndrome (Q61.9)	
Q01.0	Frontal encephalocele Encephalocystocele, frontal Encephalomyocele, frontal Hydroencephalocele, frontal Hydromeningocele, frontal Meningocele, frontal Meningoencephalocele, frontal	742.0
Q01.1	Nasofrontal encephalocele Encephalocystocele, nasofrontal Encephalomyocele, nasofrontal Hydroencephalocele, nasofrontal Hydromeningocele, nasofrontal Meningocele, nasofrontal Meningoencephalocele, nasofrontal	742.0
Q01.2	Occipital encephalocele Encephalocystocele, occipital Encephalomyocele, occipital Hydroencephalocele, occipital Hydromeningocele, occipital Meningocele, occipital Menigoencephalocele, occipital	742.0
Q01.8	Encephalocele of other sites: cranial, cerebral, nasal, parietal, other specified site (must specify) Encephalocystocele (cranial, cerebral, nasal, parietal or other specified site) Encephalomyocele (cranial, cerebral, nasal, parietal or other specified site) Hydroencephalocele (cranial, cerebral, nasal, parietal or other specified site) Hydromeningocele (cranial, cerebral, nasal, parietal or other specified site) Meningocele (cranial, cerebral, nasal, parietal or other specified site) Menigoencephalocele (cranial, cerebral, nasal, parietal or other specified site)	742.0
Q01.9	Encephalocele, unspecified (try not to use this code) Arnold-Chiari syndrome, type 3 Congenital cerebral meningocele Encephalocystocele, unspecified Encephalomyocele, unspecified Hydroencephalocele, unspecified Hydromeningocele, cranial, unspecified Meningocele, cerebral, unspecified	742.0

	Meningoencephalocele, unspecified	
Q02	<p>Microcephaly Code first, if applicable, congenital Zika virus disease Includes: hydromicrocephaly, micrencephalon Excludes 1: Meckel-Gruber syndrome (Q61.9)</p>	
Q02	<p>Microcephaly Congenital brain hypoplasia Galloway syndrome Hydromicrocephaly Micrencephalon Micrencephaly Microcephalus Primary microcephaly Use Additional Code, if applicable, to identify congenital Zika virus disease</p>	742.1
Q03	<p>Congenital hydrocephalus Excludes 1: Arnold-Chiari syndrome, type II (Q07.0-), acquired hydrocephalus (G91.-), hydrocephalus due to congenital toxoplasmosis (P37.1), hydrocephalus with spina bifida (Q05.0-Q05.4)</p>	
Q03.0	<p>Malformations of aqueduct of Sylvius (must specify) Obstruction of aqueduct of Sylvius, congenital Stenosis of aqueduct of Sylvius</p>	742.3
Q03.1	<p>Atresia of foramina of Magendie and Luschka Dandy-Walker syndrome</p>	742.3
Q03.8	<p>Other congenital hydrocephalus (must specify) Communicating hydrocephaly/enlarged Cisterna Magna Other specified hydrocephaly (do not code if Intraventricular hemorrhage/bleed) Ventriculomegaly of the Head (verify not ventriculomegaly of the heart, do not report/code if Mild dilatation)</p>	742.3
Q03.9	<p>Congenital hydrocephalus, unspecified Unspecified hydrocephaly, NOS</p>	742.3
Q04	<p>Other congenital malformations of the brain Excludes 1: cyclopia (Q87.0), macrocephaly (Q75.3)</p>	
Q04.0	<p>Congenital malformations of corpus callosum (must specify) Agenesis of corpus callosum Aplasia corpus callosum Hypoplasia, hypoplastic corpus callosum Other anomalies of the corpus callosum Partial agenesis of the corpus callosum Absent corpus callosum Congenital malformation of corpus callosum Corpus callosum anomaly, congenital Hypogenesis of corpus callosum</p>	742.2
Q04.1	<p>Arhinencephaly Arhinia</p>	742.2
Q04.2	<p>Holoprosencephaly Holoprosencephaly sequence</p>	742.2
Q04.3	<p>Other reduction deformities of the brain (must specify) Absence of part of brain Agenesis of part brain Agyria Aplasia of part of brain Hydranencephaly Hypoplasia of part of brain Lissencephaly Microgyria Pachygyria Polymicrogyria</p>	742.2

	Unspecified reduction defect of brain (try not to use) Excludes 1: congenital malformations of corpus callosum (Q04.0)	
Q04.4	Septo-optic dysplasia of brain Septo-optic dysplasia sequence <i>De Morsier syndrome</i>	742.4
Q04.5	Megalencephaly Enlarged brain and/or head Macrocephaly Macroencephaly	742.4
Q04.6	Congenital cerebral cyst(s) Colloid cyst of third ventricle Congenital pseudoporencephaly Porencephaly Schizencephaly (m) <i>Congenital choroid plexus cyst</i> <i>Porencephalic cysts</i> Excludes 1: acquired porencephalic cyst (G93.0)	742.4
Q04.8	Other specified congenital malformation of brain (must specify) Arnold-Chiari syndrome, type IV Cerebral ventriculomegaly Macrogyria Anomalies of cerebellum, includes atrophy Anomalies of hypothalamus Colpocephaly Cortical atrophy Cranial nerve defects Cystic encephalomalacia Encephalomalacia Other specified anomalies of brain Small brain Ulegyria Ventricular cysts	742.4
Q04.9	Congenital malformation of brain, unspecified (try not to use this code) Brain anomaly Congenital disease or lesion NOS of brain Multiple anomalies NOS of brain, congenital	742.9
Q05	Spina bifida (SB) Includes: hydromeningocele (spinal), meningocele (spinal), meningomyelocele, myelocele, myelomeningocele, rachischisis, spina bifida (aperta)(cystica), syringomyelocele Excludes 1: Arnold-Chiari syndrome, type II (Q07.0-), spina bifida occulta (Q76.0)	
Q05.0	Cervical spina bifida (SB) with hydrocephalus Cervical SB with hydrocephalus and hydromeningocele Cervical SB with hydrocephalus and meningocele Cervical SB with hydrocephalus and meningomyelocele Cervical SB with hydrocephalus and myelocele Cervical SB with hydrocephalus and myelomeningocele Cervical SB with hydrocephalus and rachischisis Cervical SB with hydrocephalus and syringohydromyelia Cervical SB with hydrocephalus and lipomeningomyelocele Cervical SB with hydrocephalus and lipomyelomeningocele Cervical SB with hydrocephalus and lipomeningocele Cervical SB with hydrocephalus and spina bifida (aperta)(cystica)	741.01
Q05.1	Thoracic spina bifida (SB) with hydrocephalus Dorsal spina bifida Thoracolumbar spina bifida (SB) with hydrocephalus Thoracic/dorsal/thoracolumbar SB with hydrocephalus and hydromeningocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and meningocele	741.02

	<p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and meningomyelocele</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and myelocele</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and myelomeningocele</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and rachischisis</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and syringohydromyelia</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and lipomeningomyelocele</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and lipomyelomeningocele</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and lipomeningocele</p> <p>Thoracic/dorsal/thoracolumbar SB with hydrocephalus and SB (aperta)(cystica)</p>	
<u>Q05.2</u>	<p>Lumbar spina bifida (SB) with hydrocephalus</p> <p>Lumbosacral spina bifida (SB) with hydrocephalus</p> <p>Lumbar/lumbosacral SB with hydrocephalus and hydromeningocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and meningocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and meningomyelocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and myelocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and myelomeningocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and rachischisis</p> <p>Lumbar/lumbosacral SB with hydrocephalus and syringohydromyelia</p> <p>Lumbar/lumbosacral SB with hydrocephalus and lipomeningomyelocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and lipomyelomeningocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and lipomeningocele</p> <p>Lumbar/lumbosacral SB with hydrocephalus and SB (aperta)(cystica)</p>	741.03
<u>Q05.3</u>	<p>Sacral spina bifida (SB) with hydrocephalus</p> <p>Sacral SB with hydrocephalus and hydromeningocele</p> <p>Sacral SB with hydrocephalus and meningocele</p> <p>Sacral SB with hydrocephalus and meningomyelocele</p> <p>Sacral SB with hydrocephalus and myelocele</p> <p>Sacral SB with hydrocephalus and myelomeningocele</p> <p>Sacral SB with hydrocephalus and rachischisis</p> <p>Sacral SB with hydrocephalus and syringohydromyelia</p> <p>Sacral SB with hydrocephalus and lipomeningomyelocele</p> <p>Sacral SB with hydrocephalus and lipomyelomeningocele</p> <p>Sacral SB with hydrocephalus and lipomeningocele</p> <p>Sacral SB with hydrocephalus and SB (aperta)(cystica)</p>	741.03
<u>Q05.4</u>	<p>Unspecified spina bifida (SB) with hydrocephalus</p> <p>Unspecified SB with hydrocephalus and hydromeningocele</p> <p>Unspecified SB with hydrocephalus and meningocele</p> <p>Unspecified SB with hydrocephalus and meningomyelocele</p> <p>Unspecified SB with hydrocephalus and myelocele</p> <p>Unspecified SB with hydrocephalus and myelomeningocele</p> <p>Unspecified SB with hydrocephalus and rachischisis</p> <p>Unspecified SB with hydrocephalus and syringohydromyelia</p> <p>Unspecified SB with hydrocephalus and lipomeningomyelocele</p> <p>Unspecified SB with hydrocephalus and lipomyelomeningocele</p> <p>Unspecified SB with hydrocephalus and lipomeningocele</p> <p>Unspecified SB with hydrocephalus and SB (aperta)(cystica)</p>	741.00
<u>Q05.5</u>	<p>Cervical spina bifida (SB) without hydrocephalus</p> <p>Cervical SB without hydrocephalus, with hydromeningocele</p> <p>Cervical SB without hydrocephalus, with lipomeningomyelocele</p> <p>Cervical SB without hydrocephalus, with lipomyelomeningocele</p> <p>Cervical SB without hydrocephalus, with lipomeningocele</p> <p>Cervical SB without hydrocephalus, with meningocele</p> <p>Cervical SB without hydrocephalus, with meningomyelocele</p> <p>Cervical SB without hydrocephalus, with myelocele</p> <p>Cervical SB without hydrocephalus, with myelomeningocele</p> <p>Cervical SB without hydrocephalus, with rachischisis</p> <p>Cervical SB without hydrocephalus, with SB (aperta)(cystica)</p>	741.91

	Cervical SB without hydrocephalus, with syringohydromyelia	
<u>Q05.6</u>	Thoracic spina bifida (SB) without hydrocephalus Dorsal spina bifida (SB), NOS Thoracolumbar spina bifida(SB), NOS Thoracic/dorsal/thoracolumbar SB without hydrocephaly with hydromeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with lipomeningomyelocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with lipomyelomeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with lipomeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with meningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with meningomyelocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with myelocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with myelomeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with rachischisis Thoracic/dorsal/thoracolumbar SB without hydrocephaly with SB (aperta)(cystica) Thoracic/dorsal/thoracolumbar SB without hydrocephaly with syringohydromyelia	741.92
<u>Q05.7</u>	Lumbar spina bifida (SB) without hydrocephalus Lumbosacral SB, NOS Lumbar/lumbosacral SB without hydrocephalus, with hydromeningocele Lumbar/lumbosacral SB without hydrocephalus, with lipomeningomyelocele Lumbar/lumbosacral SB without hydrocephalus, with lipomyelomeningocele Lumbar/lumbosacral SB without hydrocephalus, with lipomeningocele Lumbar/lumbosacral SB without hydrocephalus, with meningocele Lumbar/lumbosacral SB without hydrocephalus, with meningomyelocele Lumbar/lumbosacral SB without hydrocephalus, with myelocele Lumbar/lumbosacral SB without hydrocephalus, with myelomeningocele Lumbar/lumbosacral SB without hydrocephalus, with rachischisis Lumbar/lumbosacral SB without hydrocephalus, with SB (aperta)(cystica) Lumbar/lumbosacral SB without hydrocephalus, with syringohydromyelia	741.93
<u>Q05.8</u>	Sacral spina bifida (SB) without hydrocephalus Sacral SB without hydrocephalus, with hydromeningocele Sacral SB without hydrocephalus, with lipomeningomyelocele Sacral SB without hydrocephalus, with lipomyelomeningocele Sacral SB without hydrocephalus, with lipomeningocele Sacral SB without hydrocephalus, with meningocele Sacral SB without hydrocephalus, with meningomyelocele Sacral SB without hydrocephalus, with myelocele Sacral SB without hydrocephalus, with myelomeningocele Sacral SB without hydrocephalus, with rachischisis Sacral SB without hydrocephalus, with spina bifida (aperta)(cystica) Sacral SB without hydrocephalus, with syringohydromyelia	741.90
<u>Q05.9</u>	Spina bifida (SB) unspecified (without hydrocephalus) Congenital meningocele Decubitus ulcer due to spina bifida Dysraphism, congenital Hydromeningocele Hydromeningomyelocele Hydromyelocele Lipoma of spina bifida Lipomyelomeningocele Meningomyelocele Myelocele Myelomeningocele Myelomeningocele without hydrocephalus Neurogenic bladder due to spina bifida Pressure ulcer due to spina bifida Rachischisis SB without hydrocephalus	741.90

	Unspecified SB without hydrocephalus, with lipomeningomyelocele Unspecified SB without hydrocephalus, with lipomeningocele Unspecified SB without hydrocephalus, with meningocele Unspecified SB without hydrocephalus, with SB (aperta)(cystica) Unspecified SB without hydrocephalus, with syringohydromyelia	
<u>Q06</u>	Other congenital malformations of spinal cord	
<u>Q06.0</u>	Amyelia (congenital absence of spinal cord)	742.59
<u>Q06.1</u>	Hypoplasia or dysplasia of spinal cord Atelomyelia (spinal cord defect) Myelatelia Myelodysplasia of spinal cord	742.59
<u>Q06.2</u>	Diastematomyelia	742.51
<u>Q06.3</u>	Other congenital cauda equina malformations	742.59
<u>Q06.4</u>	Hydromyelia Hydrorachis	742.53
<u>Q06.8</u>	Other specified congenital malformations of spinal cord (must specify) Lumbosacral dermal sinus tract Occult spinal dysraphism sequence Tethered cord syndrome, congenital Tethered cord, filum Low lying spinal chord Tethered filum terminale <i>(m) Fibrolipoma of filum terminale</i>	742.59
<u>Q06.9</u>	Congenital malformation of spinal cord, unspecified Cervical spine dysraphism Congenital anomaly NOS of spinal cord Congenital deformity NOS of spinal cord Congenital disease or lesion NOS of spinal cord Spinal cord anomaly Split spinal cord	742.9
<u>Q07</u>	Other congenital malformations of nervous system Excludes 2: congenital central alveolar hypoventilation syndrome (G47.35), familial dysautonomia [Riley-Day] (G90.1), neurofibromatosis (nonmalignant) (Q85.0-)	
<u>Q07.0</u>	Arnold-Chiari syndrome (type II), without hydrocephalus Excludes 1: Arnold-Chiari syndrome, type III (Q01.-), Arnold-Chiari syndrome, type IV (Q04.8)	
<u>Q07.00</u>	Arnold-Chiari syndrome (type II) without spina bifida or hydrocephalus Arnold-Chiari type 2 without hydrocephalus	741.90
<u>Q07.01</u>	Arnold-Chiari syndrome (type II) with spina bifida (SB) Arnold-Chiari syndrome (type II) with SB and hydromeningocele Arnold-Chiari syndrome (type II) with SB and meningocele Arnold-Chiari syndrome (type II) with SB and meningomyelocele Arnold-Chiari syndrome (type II) with SB and myelocele Arnold-Chiari syndrome (type II) with SB and myelomeningocele Arnold-Chiari syndrome (type II) with SB and rachischisis Arnold-Chiari syndrome (type II) with SB and syringohydromyelia Arnold-Chiari syndrome (type II) with SB and lipomeningomyelocele Arnold-Chiari syndrome (type II) with SB and lipomyelomeningocele Arnold-Chiari syndrome (type II) with SB and lipomeningocele Arnold-Chiari syndrome (type II) with SB and SB (aperta)(cystica) Specify (cervical, thoracic, dorsal, thoracolumbar, lumbar, lumbosacral, sacral, unspecified)	741.90 741.91 741.92 741.93
<u>Q07.02</u>	Arnold-Chiari syndrome (type II) with hydrocephalus Arnold-Chiari Type 2 with hydrocephalus	742.0
<u>Q07.03</u>	Arnold-Chiari syndrome (type II) with spina bifida (SB) and hydrocephalus Arnold-Chiari syndrome (type II) with SB, hydrocephalus and hydromeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and meningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and meningomyelocele	741.00 741.01 741.02 741.03

	Arnold-Chiari syndrome (type II) with SB, hydrocephalus and myelocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and myelomeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and rachischisis Arnold-Chiari syndrome (type II) with SB, hydrocephalus and syringohydromyelia Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomeningomyelocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomyelomeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and SB (aperta)(cystica) Specify (cervical, thoracic, dorsal, thoracolumbar, lumbar, lumbosacral, sacral, unspecified)	
<u>Q07.8</u>	Other specified congenital malformations of nervous system (must specify) Agenesis of nerve Congenital optic nerve pit Displacement of brachial plexus Incomplete development of nerve Jaw-winking syndrome Marcus Gunn's syndrome Marcus Gunn jaw winking syndrome Pit of optic disc Synkinesis, eyelid	742.8
<u>Q07.9</u>	Congenital malformation of nervous system, unspecified Central nervous system (CNS) malformation, fetal Congenital anomaly NOS of nervous system Congenital deformity NOS of nervous system Congenital disease or lesion NOS of nervous system Nervous system anomaly Neutral tube defect	742.9

Congenital Malformations of Eye, Ear, Face and Neck (Q10-Q18)

ICD-10	Description	ICD-9
Q10	Congenital malformation of eyelid, lacrimal apparatus and orbit Excludes 1: cryptophthalmos NOS (Q11.2), cryptophthalmos syndrome (Q87.0)	
Q10.0	Congenital ptosis Bilateral congenital ptosis Left congenital ptosis Right congenital ptosis Blepharoptosis	743.61
Q10.1	Congenital ectropion	743.62
Q10.2	Congenital entropion	743.62
Q10.3	Other congenital malformations of eyelid (must specify) Ablepharon Blepharophimosis, congenital Coloboma of eyelid Congenital absence or agenesis of eyelid Congenital accessory eyelid Congenital accessory eye muscle Telecanthus (m) Congenital absence or agenesis of cilia (m) Congenital malformation of eyelid NOS (m) Absence of eyelashes (m) Epicantal folds (m) Fused eyelids (if birth weight greater than 1000 grams) (m) Slanted eyes (m) Small or narrow palpebral fissures (m) Upward eyeslant (m) Weakness of eyelids Do not report pseudostrabismus	743.62 743.63
Q10.4	(m) Absence and agenesis of lacrimal apparatus (m) Congenital absence of punctum lacrimal (m) Nasolacrimal duct obstruction	743.65
Q10.5	(m) Congenital stenosis and stricture of lacrimal duct (m) Congenital blocked tear duct (m) Congenital dacryostenosis (left/ right/ bilateral) (m) Obstruction of lacrimal duct(s)	743.65
Q10.6	(m) Other congenital malformations of lacrimal apparatus (m) Congenital abnormality of lacrimal drainage system (m) Congenital absence of lacrimal drainage structure/lacrimal apparatus (m) Congenital anomaly of lacrimal gland (m) Accessory lacrimal canal	743.65
Q10.7	Congenital malformation/anomaly of orbit/ orbit proper Congenital superior sulcus anomaly of orbit	743.66
Q11	Anophthalmos, microphthalmos and macropthalmos	
Q11.0	Cystic eyeball Congenital cystic globe	743.03
Q11.1	Other anophthalmos Anophthalmos, NOS Agenesis of eye Aplasia of eye Congenital absence of the eye	743.00
Q11.2	Microphthalmos (eye condition) Cryptophthalmos Dysplasia of eye Hypoplasia of eye Rudimentary eye	743.06 743.10 743.11 743.12

	Simple microphthalmos Microphthalmos associated with other anomalies of eye and adnexa Excludes 1: cryptophthalmos syndrome (Q87.0)	
Q11.3	Macrophthalmos Excludes 1: macrophtalmos in congenital glaucoma (Q15.0)	743.8
Q12	Congenital lens malformations	
Q12.0	Congenital cataract (Please specify right, left or bilateral) Anterior and posterior axial embryonal cataract Capsular and subcapsular cataract Cataract, anterior polar Cataract, NOS Cataract, other specified Combined form cataract Cortical and zonular cataract Nuclear cataract Total and subtotal cataract, congenital	743.30 743.31 743.32 743.33 743.34
Q12.1	Congenital displaced lens Congenital ectopic lens Ectopia lentis	743.37
Q12.2	Coloboma of lens	743.36
Q12.3	Congenital aphakia Absence of lens	743.35
Q12.4	Spherophakia	743.36
Q12.8	Other congenital lens malformations (must specify) Micropakia Anomalies of lens shape Other specified lens anomaly Mittendorf Dot is not reportable	743.36
Q12.9	Congenital lens malformation, unspecified Congenital anomaly of lens shape Unspecified lens anomaly	743.39
Q13	Congenital malformations of anterior segment of eye	
Q13.0	Coloboma of iris Coloboma, NOS	743.46
Q13.1	Absence of iris Aniridia	743.45
Q13.2	Other congenital malformations of iris (must specify) (m) Anisocoria, congenital (m) Anomaly of iris and/or ciliary body (m) Atresia of pupil (m) Brushfield spots (m) Congenital heterochromia iridis (m) Congenital iris heterochromia (m) Congenital pupil anomaly (m) Corectopia (m) Deformed pupil (m) Dyscoria (m) Ectopic pupil (m) Glaucoma assoc with or due to iris anomaly (m) Iridial remnants (m) Polycoria	743.46
Q13.3	Congenital corneal opacity Congenital arcus juvenilis Corneal opacities, interfering with vision, congenital	743.42
Q13.4	Other congenital corneal malformations (size and shape) (must specify) Congenital malformations of cornea, NOS Irido-corneo-trabecular dysgenesis	743.22 743.41

	Microcornea Peter's anomaly (m) Megalocornea (without glaucoma) (m) Congenital megalocornea (without glaucoma) (m) Enlarged cornea (without glaucoma)	
<u>Q13.5</u>	(m) Blue sclera (m) Scleral anomaly (m) Specified anomalies of sclera	743.47
<u>Q13.8</u>	Other congenital malformations of anterior segment of eye	
<u>Q13.81</u>	Rieger's anomaly Irido-trabecular dysgenesis Rieger eye malformation sequence	743.44
<u>Q13.89</u>	Other congenital malformations of anterior segment of eye (must specify) Multiple and combined anomaly of anterior segment Other specified colobomas and anomalies of anterior segments (m) Congenital scleral show	743.47 743.48 743.49
<u>Q13.9</u>	Congenital malformations of anterior segment of eye, unspecified Anterior segment anomaly, eye Anterior segment anomaly, multiple, eye Congenital anomaly of anterior chamber or segment of eye Glaucoma associated with or due to chamber angle anomaly Multiple anterior segment anomalies Unspecified anomalies of anterior segments or anterior chamber	743.49
<u>Q14</u>	Congenital malformations of posterior segment of eye Excludes 2: optic nerve hypoplasia (H47.03-)	
<u>Q14.0</u>	Congenital malformation of vitreous humor Congenital vitreous opacity Persistent hyperplastic primary vitreous	743.51
<u>Q14.1</u>	Congenital malformations of retina Congenital retinal aneurysm Congenital macular changes Other retinal changes, congenital Specified anomaly of retina	743.55 743.56 743.58
<u>Q14.2</u>	Congenital malformations of optic disc Coloboma of optic disc Congenital optic disc coloboma Optic nerve pit, congenital Pit of optic disc Specified anomaly of optic disc	743.57
<u>Q14.3</u>	Congenital malformation of choroid Chorioretinal degeneration, congenital Coloboma of choroid folds Congenital choroidal folds	743.53
<u>Q14.8</u>	Other congenital malformations of posterior segment of eye (must specify) Chroiretinal coloboma Coloboma of the fundus Congenital hypoplasia of fovea centralis Fovea hypoplasia Congenital folds and cysts of posterior segment Vascular anomalies	743.52 743.58
<u>Q14.9</u>	Congenital malformation of posterior segment of eye, unspecified Unspecified anomaly of posterior segment of eye	743.59
<u>Q15</u>	Other congenital malformations of eye Excludes 1: congenital nystagmus (H55.01), ocular albinism (E70.31-), optic nerve hypoplasia (H47.03-), retinitis pigmentosa (H35.52)	
<u>Q15.0</u>	Congenital glaucoma Axenfeld's anomaly Buphthalmos	743.20 743.21 743.22

	Enlarged eye, NOS Glaucoma of childhood Glaucoma of newborn Hydrophthalmos, simple, unspecified Juvenile glaucoma Keratoglobus, congenital, with glaucoma Macrocornea with glaucoma Macrophthalmos in congenital glaucoma Megalocornea with glaucoma Primary congenital glaucoma Simple buphtalmos Buphtalmos associated with other ocular anomalies Congenital glaucoma, simple	743.44
<u>Q15.8</u>	Other specified congenital malformations of eye (must specify) Epibulbar dermoid cyst	743.8
<u>Q15.9</u>	Congenital malformation of eye, unspecified (try not to use) Congenital anomaly of eye Congenital deformity of eye	743.9
<u>Q16</u>	Congenital malformations of ear causing impairment of hearing Excludes 1: congenital deafness (H90.-)	
<u>Q16.0</u>	Congenital absence of (ear) auricle (Anotia) Absence of external ear	744.01
<u>Q16.1</u>	Congenital absence, atresia or stricture of auditory canal (external) (must specify) Congenital atresia or stricture of osseous meatus Congenital atresia of external auditory canal Congenital atresia of external ear (left/right/bilateral) Aural atresia Absence of auditory canal Absence of external auditory meatus (m) Narrow external auditory meatus	744.02
<u>Q16.2</u>	Absence of Eustachian tube	744.24
<u>Q16.3</u>	Congenital malformation of ear ossicles Congenital fusion of ear ossicles	744.04
<u>Q16.4</u>	Other congenital malformations of middle ear Congenital malformations of middle ear, NOS	744.03
<u>Q16.5</u>	Congenital malformation of inner ear Congenital anomaly of membranous labyrinth Congenital anomaly of organ of Corti	744.05
<u>Q16.9</u>	Congenital malformation of ear causing impairment of hearing, unspecified Congenital absence of ear, NOS Congenital external ear anomaly with hearing impairment	744.00
<u>Q17</u>	Other congenital malformations of ear Excludes 1: congenital malformations of ear with impairment of hearing (Q16.0-Q16.9), preauricular sinus (Q18.1)	
<u>Q17.0</u>	(m) Accessory auricle of ear (m) Accessory tragus (m) Polyotia (m) Preauricular appendage or skin tag (m) Supernumerary ear (m) Supernumerary lobule (m) Ear, double lobule (m) Other appendage, tag, or lobule (m) Papillomas, ear tags (m) Skin tag of pinna	744.1
<u>Q17.1</u>	(m) Macrotia (<i>enlarged pinna</i>)	744.22
<u>Q17.2</u>	Microtia (<i>hypoplastic pinna</i>)	744.23
<u>Q17.3</u>	(m) Other misshapen ear (m) Lop ear	744.29

	(m) Pointed ear (m) Absent or decreased cartilage (m) Cleft ear (m) Crumpled ears (m) Cupped ears (m) Darwin's tubercle (m) Ears, absent tragus (m) Ears, bridged concha (m) Ears, lack of helical fold (m) Ears, thickened or over folded helix (m) Extra cartilage (m) Malformed ear (m) Notch in ear (m) Posteriorly rotated ears (m) Protruding ears	
Q17.4	(m) Misplaced ear (m) Low-set ears Excludes 1: cervical auricle (Q18.2)	744.29
Q17.5	(m) Prominent ear (m) Bat ear	744.29
Q17.8	(m) Other specified congenital malformations of ear (must specify) (m) Congenital absence of lobe of ear (m) Congenital split ear lobes (left/right/bilateral) (m) Cryptotia	744.21 744.24 743.29
Q17.9	(m) Congenital malformation of ear, unspecified (m) Congenital anomaly of ear, NOS (m) Congenital external ear (pinna) anomaly (m) Congenital anomaly of ear (left/right/bilateral)	744.3
Q18	Other congenital malformations of face and neck Excludes 1: cleft lip and cleft palate (Q35-Q37), conditions classified to Q67.0-Q67.4 , congenital malformations of skull and face bones (Q75.-), cyclopia (Q87.0), dentofacial anomalies [including malocclusion] (M26.-), malformation syndromes affecting facial appearance (Q87.0), persistent thyroglossal duct (Q89.2)	
Q18.0	(m) Sinus, fistula and cyst of branchial cleft (m) Branchial vestige (m) Branchial cleft fistula (m) Branchial cleft sinus (m) Branchial cleft cyst (m) Branchial cleft pit	744.41
Q18.1	(m) Preauricular sinus and cyst (m) Cervicoaural fistula (m) Fistula of auricle, congenital (m) Ear pit (m) Preauricular fistula (m) Preauricular pits (m) Preauricular sinus	744.46 744.47
Q18.2	(m) Other branchial cleft malformations (must specify) (m) Branchial cleft malformations, NOS (m) Cervical auricle (m) Otocephaly (m) Dermal sinus of head (m) Other branchial cleft anomaly	744.43 744.49
Q18.3	(m) Webbing of neck (m) Pterygium colli (m) Redundant neck skin folds	744.5
Q18.4	Macrostomia	744.83
Q18.5	Microstomia	744.84

<u>Q18.6</u>	(m) <i>Macrocheilia (large lips)</i> (m) <i>Hypertrophy of lip, congenital</i>	744.81
<u>Q18.7</u>	(m) <i>Microcheilia</i>	744.82
<u>Q18.8</u>	Other specified congenital malformations of face and neck (must specify) Medial cyst of face and neck Medial fistula of face and neck Medial sinus of face and neck <i>Cleft face (excluding cleft lip and palate)</i> (m) <i>Short neck</i>	744.89
<u>Q18.9</u>	Congenital malformations of face and/or neck, unspecified (try not to use) Congenital anomaly NOS of face and/or neck (m) <i>Abnormal facies</i> (m) <i>Craniofacial dysmorphosis</i> (m) <i>Dysmorphic facies/face</i>	744.89 744.9

Congenital Malformations of the Circulatory System (Q20-Q28)

ICD-10	Descriptions	ICD-9
Q20	Congenital malformations of cardiac chambers and connections Excludes 1: dextrocardia with situs inversus (Q89.3), mirror-image atrial arrangement with situs inversus (Q89.3)	
Q20.0	Common arterial trunk Persistent truncus arteriosus <i>Absent septum between aorta and pulmonary artery</i> <i>Common aortopulmonary trunk</i> Excludes 1: aortic septal defect (Q21.4)	745.0
Q20.1	Double outlet right ventricle Taussig-Bing syndrome	745.11
Q20.2	Double outlet left ventricle	745.19
Q20.3	Discordant ventriculoarterial connection Dextraposition of aorta Dextro-transposition of aorta Dextroposition of aorta Dextrotransposition of the great arteries Incomplete great vessel transposition Incomplete transposition of the great vessels L-transposition of the great vessels Levotransposition of the great arteries Transposition of great vessels (complete) D-transposition	745.10
Q20.4	Double inlet ventricle (right or left) Common ventricle Cor triloculare biventriculatum Single ventricle	745.3
Q20.5	Discordant atrioventricular connection Corrected transposition Levo-transposition Ventricular inversion L-transposition, ventricular inversion <i>Other specific transposition of great vessels</i> <i>Unspecified transposition of great vessels</i>	745.12
Q20.6	Isomerism of atrial appendages Isomerism of atrial appendages with asplenia or polysplenia	745.8 759.0
Q20.8	Other congenital malformations of cardiac chambers and connections (must specify) Cor binoculare <i>Absence of atrial and ventricular septa</i> <i>Two chambered heart</i>	745.19 745.7 745.8
Q20.9	Congenital malformation of cardiac chambers and connections, unspecified	746.9
Q21	Congenital malformations of cardiac septa Excludes 1: acquired cardiac septal defect (I51.0)	
Q21.0	Ventricular septal defect (VSD) Roger's disease <i>VSD, Malalignment (type I, subarterial, subaortic, subpulmonic; outlet, supracristal)</i> <i>VSD, Perimembranous (type II, membranous, infracristal, subaortic, Gerbode)</i> <i>VSD, Endocardial cushion defect (type III; inlet, inflow type, subtricuspid, canal-type)</i> <i>VSD, Muscular (type IV; trabecular, swiss cheese, central, apical, marginal, Roger's disease)</i> Interventricular septal defect	745.4

<u>Q21.1</u>	Atrial septal defect (ASD) Coronary sinus defect Patent or persistent ostium secundum defect (type II) <u>Atrial septal defect, NOS</u> <u>Auricular septal defect, NOS</u> <u>Secundum atrial septal defect</u> <i>(m) Patent foramen ovale (PFO)</i> <i>(m) Patent or persistent sinus venosus defect</i> <i>(m) Persistent foramen ovale (PFO)</i> <i>(m) Fossa ovalis defect</i> <i>(m) Foramen ovale</i> <i>(m) Nonclosure of foramen ovale, NOS</i> <i>(m) Partial foramen ovale</i>	745.5
<u>Q21.2</u>	Atrioventricular septal defect (AVSD) Atrial septal defect, primum Common atrioventricular canal Endocardial cushion defect Ostium primum atrial septal defect (ASD) (type I) Ostium primum defect <u>Absence of atrial septum</u> <u>Artrioventricular canal type VSD</u> <u>Common atrioventricular canal with VSD</u> <u>Other endocardial cushion defects</u> <u>Persistent ostium primum</u> <u>Single common atrium</u> <u>Type III VSD, Canal-type VSD</u>	745.60 745.61 745.69
<u>Q21.3</u>	Tetralogy of Fallot (ventricular septal defect with pulmonary stenosis or atresia, dextroposition of aorta and hypertrophy of right ventricle) (also specify all individual defects) Dextraposition of aorta in Fallot's tetralogy Tetralogy of Fallot with absent pulmonary valve Tetralogy of Fallot with pulmonary atresia or stenosis Tetralogy of Fallot with ventricular septal defect	745.2
<u>Q21.4</u>	Aortopulmonary septal defect Abnormal communication between aorta and pulmonary artery Aortic septal defect Aortopulmonary window	745.0
<u>Q21.8</u>	Other congenital malformations of cardiac septa (must specify) Eisenmenger's defect Pentalogy of Fallot <u>Fallot's tetralogy plus atrial septal defect</u>	745.8 745.2
<u>Q21.9</u>	Congenital malformations of cardiac septum, unspecified Septal (heart) defect, NOS	745.9
<u>Q22</u>	Congenital malformations of pulmonary and tricuspid valves	
<u>Q22.0</u>	Pulmonary valve atresia Congenital absence of pulmonary valve	746.01
<u>Q22.1</u>	Congenital pulmonary valve stenosis (narrowing)	746.02
<u>Q22.2</u>	Congenital pulmonary valve insufficiency (do not code if mild, trivial or physiologic) Congenital pulmonary valve regurgitation (do not code if mild, trivial or physiologic) <u>Trilogy of Fallot</u>	746.09
<u>Q22.3</u>	Other congenital malformations of pulmonary valve (must specify) Congenital malformation of pulmonary valve, NOS Supernumerary cusps of pulmonary valve <u>Bicuspid pulmonary valve</u> <u>Other specified anomaly of pulmonary valve</u> <u>Pulmonary valve hypoplasia</u> <u>Quadracuspid pulmonary valve</u>	746.00 746.09

Q22.4	Congenital tricuspid valve stenosis (narrowing) Congenital tricuspid atresia Absence of tricuspid valve Tricuspid hypoplasia	746.1
Q22.5	Ebstein's anomaly Ebstein's anomaly of tricuspid valve	746.2
Q22.6	Hypoplastic right heart syndrome	756.89
Q22.8	Other congenital malformations of tricuspid valve (must specify) Insufficiency of tricuspid valve Congenital tricuspid valve regurgitation	746.89
Q22.9	Congenital malformations of tricuspid valve, unspecified	746.89
Q23	Congenital malformations of aortic and mitral valves	
Q23.0	Congenital stenosis of aortic valve Congenital aortic atresia Congenital aortic stenosis, NOS Type 1 Excludes: Congenital stenosis of aortic valve in hypoplastic left heart syndrome (Q23.4), congenital subaortic stenosis (Q24.4), supravalvular aortic stenosis (congenital) (Q25.3)	746.3
Q23.1	Congenital insufficiency of aortic valve (do not code if mild, trivial or physiologic) Bicuspid aortic valve Congenital aortic insufficiency Congenital aortic valve regurgitation	746.4
Q23.2	Congenital mitral stenosis (narrowing) Congenital mitral atresia Congenital supravalvular mitral stenosis Absence of mitral valve Atresia of mitral valve Hypoplasia of mitral valve/cleft mitral valve	746.5
Q23.3	Congenital mitral insufficiency (do not code if mild, trivial or physiologic) Congenital mitral valve regurgitation Mitral valve prolapse	746.6
Q23.4	Hypoplastic left heart syndrome	746.7
Q23.8	Other congenital malformations of aortic and mitral valves (must specify) Cleft leaflet of mitral valve Aortic valve atresia/absence Fused commissure of mitral valve Other specified anomaly of aortic valves Parachute deformity of mitral valve Supernumerary cusps	746.89
Q23.9	Congenital malformation of aortic and mitral valves, unspecified Unspecified anomaly of the aortic valves	746.89
Q24	Other congenital malformations of heart Excludes 1: endocardial fibroelastosis (I42.4)	
Q24.0	Dextrocardia Dextrocardia with no mention of situs inversus Dextrocardia without situs inversus (situs solitus) Excludes 1: dextrocardia with situs inversus (Q89.3), isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6), mirror-image atrial arrangement with situs inversus (Q89.3)	746.87
Q24.1	Levocardia Situs inversus with levocardia	746.87
Q24.2	Cor triatriatum Cor triatriatum, dexter/left sided/right sided/sinister Divided left atrium Divided right atrium	746.82
Q24.3	Pulmonary infundibular stenosis (narrowing) Infundibular pulmonic stenosis Stenosis of infundibulum of right ventricle	746.83

	Subvalvular pulmonic stenosis	
Q24.4	Congenital subaortic stenosis Congenital subvalvular aortic stenosis	746.81
Q24.5	Malformation of coronary vessels Congenital coronary (artery) aneurysm Congenital coronary artery fistula Absence of coronary artery Anomalies of coronary artery or sinus Anomalous origin or communication of coronary artery Coronary artery arising from aorta or pulmonary trunk Single coronary artery	746.85
Q24.6	Congenital heart block Complete or incomplete atrioventricular (AV) block	746.86
Q24.8	Other specified congenital malformations of heart (must specify) Congenital diverticulum of left ventricle Congenital malformation of myocardium Congenital malformation of pericardium Malposition of heart Uhl's disease (hypoplastic right ventricle) Abdominal heart Absence of papillary muscle of luthisis Anomalies of myocardium Anomalies of pericardium Anomaly of papillary muscle Atresia of cardiac vein Congenital pericardial defect Ectopia (ectopic) cordis (mesocardia) Hypoplasia of cardiac vein Hypoplastic left ventricle Hypoplastic ventricle, NOS Obstructive anomalies of heart, NEC Conduction defects, NOS Other defects of the atria Other specified anomaly of heart Shones syndrome (Complex) = 1) Parachute Mitral Valve; 2) supravalvular ring of L atrium; 3) subaortic stenosis; 4) coarctation of aorta [Code the stenosis and coarctation separately] Ventricular hypertrophy (right or left) <i>(m) Congenital cardiomegaly</i> <i>Cardiomyopathy, hypertrophic is not reportable</i>	746.84 746.87 746.89
Q24.9	Congenital malformations of heart, unspecified (try not to use) Complex congenital heart defect (code component defects) Congenital anomaly of heart Congenital disease of heart	746.9
Q25	Congenital malformations of great arteries	
Q25.0	Patent ductus arteriosus (PDA) (do not report if birth weight <1500 grams) Patent ductus Botallo Persistent ductus arteriosus Patent ductus arteriosis closed with medication	747.0
Q25.1	Coarctation of aorta Coarctation of aorta (preductal) (postductal) Unspecified coarctation of aorta	747.10
Q25.2	Atresia of aorta	
Q25.21	Interruption of aortic arch Atresia of aortic arch Interrupted aortic arch Interrupted aortic arch. Type A	747.11

	Interrupted aortic arch. Type B	
Q25.29	Other atresia of aorta Atresia of aorta	747.22
Q25.3	Supravalvular aortic stenosis (narrowing) Small aorta, narrowing of aorta Stenosis of aorta Stricture of aorta Excludes 1: congenital aortic stenosis NOS (Q23.0); congenital stenosis of aortic valve (Q23.0)	747.22
Q25.4	Other congenital malformations of aorta (must specify) Double aortic arch (Vascular ring of aorta) Hypoplasia of aorta Hypoplasia of aortic arch, congenital Hypoplastic aorta syndrome Kommerells diverticulum Overriding aorta Persistent right aortic arch Pseudocoarctation of aorta Right aortic arch <i>Aberrant subclavian artery</i> <i>Anomalous origin, right subclavian artery</i> <i>Dextroposition of aorta</i> <i>Hypoplastic aortic isthmus</i> <i>Pseudotruncus arteriosus</i> <i>Tubular hypoplasia of aorta</i> <i>Vascular ring compression of trachea</i> Excludes 1: hypoplasia of aorta in hypoplastic left heart syndrome (Q23.4)	
Q25.40	(m) <i>Congenital malformation of aorta unspecified</i>	747.20
Q25.41	Absence and aplasia of aorta	747.29
Q25.42	Hypoplasia of aorta	747.29
Q25.43	Congenital aneurysm of aorta Congenital aneurysm of aortic root Congenital aneurysm of aortic sinus	747.29
Q25.44	(m) <i>Congenital dilation of aorta</i>	747.29
Q25.45	Double aortic arch Vascular ring of aorta	747.21
Q25.46	(m) <i>Tortuous aortic arch</i> (m) <i>Persistent convolutions of aortic arch</i>	747.21
Q25.47	(m) <i>Right aortic arch</i> (m) <i>Persistent right aortic arch</i>	747.21
Q25.48	(m) <i>Anomalous origin of subclavian artery</i>	747.29
Q25.49	(m) <i>Other congenital malformations of aorta (must specify)</i> (m) <i>Aortic arch</i> (m) <i>Bovine arch</i>	747.29
Q25.5	Atresia of pulmonary artery Pulmonary artery absence Pulmonary artery agenesis Pulmonary artery atresia	747.31
Q25.6	Stenosis of pulmonary artery Supravalvular pulmonary stenosis (narrowing) <i>Peripheral pulmonary artery stenosis (do not report if birth weight <2500 g)</i> <i>Peripheral pulmonic stenosis (PPS) (do not report if birth weight <2500 g)</i>	747.31 747.39
Q25.7	Other congenital malformations of pulmonary artery	
Q25.71	Coarctation of pulmonary artery	747.31
Q25.72	Congenital pulmonary arteriovenous malformation Congenital pulmonary arteriovenous aneurysm <i>Aneurysm of pulmonary artery</i>	747.32

Q25.79	Other congenital malformations of pulmonary artery (must specify) Aberrant pulmonary artery Agenesis of pulmonary artery Congenital aneurysm of pulmonary artery Congenital anomaly of pulmonary artery Hypoplasia of pulmonary artery Dilatation of pulmonary artery Unspecified anomaly of pulmonary artery	747.39
Q25.8	Other congenital malformations of other great arteries Anomalous origin of right subclavian artery	747.29
Q25.9	Congenital malformations of great arteries, unspecified	747.29
Q26	Congenital malformations of great veins	
Q26.0	Congenital stenosis of vena cava Congenital stenosis of vena cava (inferior or superior)	747.49
Q26.1	Persistent left superior vena cava	747.49
Q26.2	Total anomalous pulmonary venous connection Total anomalous pulmonary venous return [TAPVR], subdiaphragmatic Total anomalous pulmonary venous return [TAPVR], supradiaphragmatic Total anomalous lung veins to coronary sinus/liver veins/right heart/vena cavae	747.41
Q26.3	Partial anomalous pulmonary venous connection Partial anomalous pulmonary venous return [PAPVR]	747.42
Q26.4	Anomalous pulmonary venous connection, unspecified	747.42
Q26.5	Anomalous portal venous connection	747.61
Q26.6	Portal vein-hepatic artery fistula	747.61
Q26.8	Other congenital malformations of great veins (must specify) Absence of vena cava (inferior or superior) Atresia of vena cava (inferior or superior) Azygos continuation of inferior vena cava Persistent left posterior cardinal vein Pulmonary vein stenosis Scimitar syndrome Stenosis of vena cava (inferior or superior) Anomalous portal vein termination Transposition of pulmonary veins, NOS	747.49
Q26.9	Congenital malformation of great vein, unspecified Congenital anomaly of vena cava (inferior) (superior), NOS Anomaly NOS of pulmonary veins	747.40
Q27	Other congenital malformations of peripheral vascular system Excludes 2: anomalies of cerebral and precerebral vessels (Q28.0-Q28.3), anomalies of coronary vessels (Q24.5), anomalies of pulmonary artery (Q25.5-Q25.7), congenital retinal aneurysm (Q14.1), hemangioma and lymphangioma (D18.-)	
Q27.0	(m) <i>Congenital absence and hypoplasia of umbilical artery</i> (m) <i>Single umbilical artery</i> (m) <i>Two vessels in umbilical cord</i>	747.5
Q27.1	Congenital renal artery stenosis	747.62
Q27.2	Other congenital malformations of renal artery (must specify) Congenital malformation of renal artery, NOS Multiple renal arteries Renal vessel anomaly	747.62
Q27.3	Arteriovenous malformation (AVM) (peripheral) Arteriovenous aneurysm	
Q27.30	Arteriovenous malformation, site unspecified Peripheral arteriovenous malformation	747.69
Q27.31	Arteriovenous malformation of vessel of upper limb	747.63
Q27.32	Arteriovenous malformation of vessel of lower limb	747.64
Q27.33	Arteriovenous malformation of digestive system vessel	747.61

	Arteriovenous malformation of duodenum and jejunum Arteriovenous malformation of intestinal vessels Congenital arteriovenous malformation of the gastrointestinal tract Congenital duodenal and jejunal arteriovenous malformation	
Q27.34	Arteriovenous malformation of renal vessel	747.62
Q27.39	Arteriovenous malformation, other site Arteriovenous malformation of spine	747.69
Q27.4	Congenital phlebectasia	747.89
Q27.8	Other specified congenital malformations of peripheral vascular system (must specify) Absence of peripheral vascular system Anomalous origin of right subclavian artery Atresia of peripheral vascular system Congenital aneurysm (peripheral) Congenital stricture, artery Congenital varix <i>Absence of artery, NEC</i> Anomaly of spinal vessel Atresia of artery Gastrointestinal vessel anomaly Other anomaly of peripheral arteries Other anomaly of peripheral veins Excludes 1: arteriovenous malformation (Q27.3-)	747.69
Q27.9	Congenital malformation of peripheral vascular system, unspecified Anomaly of artery or vein, NOS Arteriovenous malformation Congenital anomaly of peripheral blood vessel Congenital vascular malformation of orbit Vascular or venous malformation	747.60
Q28	Other congenital malformations of circulatory system Excludes 1: congenital aneurysm NOS (Q27.8), congenital coronary aneurysm (Q24.5), ruptured cerebral arteriovenous malformation (I60.8), ruptured malformation of precerebral vessels (I72.0) Excludes 2: congenital peripheral aneurysm (Q27.8), congenital pulmonary aneurysm (Q25.79), congenital retinal aneurysm (Q14.1)	
Q28.0	Arteriovenous malformation of precerebral vessels Congenital arteriovenous precerebral aneurysm (non-ruptured)	747.89
Q28.1	Congenital malformation of precerebral vessels, NOS Congenital precerebral aneurysm (nonruptured)	747.89
Q28.2	Arteriovenous malformation of cerebral vessels Arteriovenous malformation of brain, NOS Congenital arteriovenous cerebral aneurysm (nonruptured) Cerebral arteriovenous malformation <i>Anomalies of cerebrovascular system</i> <i>Anomalies of vein of Galen</i> <i>Other anomaly of cerebral vessels</i>	747.81
Q28.3	Other malformations of cerebral vessels (must specify) Congenital cerebral aneurysm (nonruptured) Congenital malformation of cerebral vessels, NOS Developmental venous anomaly	747.81
Q28.8	Other specified congenital malformations of circulatory system (must specify) Congenital aneurysm, specified site NEC Spinal vessel anomaly	747.89 747.82
Q28.9	Congenital malformation of circulatory system, unspecified	747.9

Congenital Malformations of the Respiratory System (Q30-Q34)

ICD-10	Description	ICD-9
Q30	Congenital malformations of nose Excludes 1: congenital deviation of nasal septum (Q67.4)	
Q30.0	Choanal atresia Atresia of nares (anterior) (posterior) Congenital stenosis of nares (anterior) (posterior) Choanal stenosis	748.0
Q30.1	Agenesis and underdevelopment of nose Agenesis of nose Congenital absent of nose	748.1
Q30.2	Fissured, notched and cleft nose Cleft nose anomaly Congenital cleft nose Congenital cleft nose anomaly Midline fissured, notched and cleft nose	748.1
Q30.3	(m) <i>Congenital perforated nasal septum</i>	748.1
Q30.8	(m) <i>Other congenital malformations of nose (must specify)</i> (m) <i>Accessory nose</i> (m) <i>Congenital anomaly of nasal sinus wall</i> (m) <i>Congenital malformation of nasal septum (nasal septal defect)</i> (m) Absent nasal septum (m) Anteverted nares (m) Flat or wide nasal bridge or other minor nose malformation (m) Notched or hypoplastic alae nasi (m) Other anomalies of nose (m) Small nares (m) Small nose and nostril (m) Tubular nose, single nostril, proboscis (m) Upturned nose (m) Wide nasal bridge	748.1
Q30.9	(m) <i>Congenital malformation of nose, unspecified</i> (m) <i>Congenital anomaly of the nose</i>	748.1
Q31	Congenital malformation of larynx Excludes 1: congenital laryngeal stridor, NOS (P28.89)	
Q31.0	Web of larynx Glottic web of larynx Laryngeal web Subglottic web Subglottic web of larynx	748.2
Q31.1	Congenital subglottic stenosis	748.3
Q31.2	Laryngeal hypoplasia	748.3
Q31.3	(m) <i>Laryngocèle</i>	748.3
Q31.5	(m) <i>Congenital laryngomalacia</i>	748.3
Q31.8	Other congenital malformations of larynx <i>(must specify)</i> Absence of larynx Agenesis of larynx Atresia of larynx Congenital cleft larynx Congenital cleft thyroid cartilage Congenital fissure of epiglottis Congenital stenosis of larynx, NEC Laryngeal cleft Posterior cleft of cricoid cartilage Anomalies of larynx and supporting cartilage Anomaly of epiglottis Anomaly of thyroid cartilage	748.3

	Atresia of epiglottis Atresia glottis Cleft larynx, laryngotracheoesophageal cleft Cleft thyroid, cartilage, congenital	
<u>Q31.9</u>	Congenital malformation of larynx, unspecified Congenital laryngeal anomaly	748.3
<u>Q32</u>	Congenital malformations of trachea and bronchus Excludes 1: congenital bronchiectasis (Q33.4)	
<u>Q32.0</u>	(m) <i>Congenital tracheomalacia</i>	748.3
<u>Q32.1</u>	Other congenital malformations of trachea (must specify) Atresia of trachea Congenital anomaly of tracheal cartilage Congenital dilation of trachea Congenital malformation of trachea Congenital stenosis of trachea Congenital tracheocele Congenital laryngotracheal anomaly Diverticulum of trachea Rudimentary tracheal bronchus	748.3
<u>Q32.2</u>	Congenital bronchomalacia	748.3
<u>Q32.3</u>	Congenital stenosis of bronchus	748.3
<u>Q32.4</u>	Other congenital malformations of bronchus (must specify) Absence of bronchus Agenesis of bronchus Atresia of bronchus Congenital diverticulum of bronchus Congenital malformations of bronchus, NOS	748.3
<u>Q33</u>	Congenital malformations of lung	
<u>Q33.0</u>	Congenital cystic lung Bronchogenic cyst Congenital cystic adenomatoid malformation of lung Congenital cystic lung disease Congenital honeycomb lung Congenital polycystic lung disease Single lung cyst CCAM (congenital cystic adenomatoid malformation) Cystic adenomatoid dysplasia of lung Cystic adenomatoid lung Cystic lung, NOS Lobar adenomatosis Lung cyst, NOS Multiple cysts, lung Other specified congenital cystic lung Polycystic lung (do not use this code for broncopulmonary dysplasia) Excludes 1: Cystic Fibrosis (E84.0), cystic lung disease, acquired or unspecified (J98.4)	748.4
<u>Q33.1</u>	Accessory lobe of lung Accessory lung Azygos lobe (fissured), lung	748.69
<u>Q33.2</u>	Sequestration of lung Congenital sequestration of lung Pulmonary sequestration	748.5
<u>Q33.3</u>	Agenesis of lung Congenital absence of lung (lobe) Congenital absence of lung	748.5
<u>Q33.4</u>	Congenital bronchiectasis	748.61
<u>Q33.5</u>	Ectopic tissue in lung	748.69

<u>Q33.6</u>	Congenital hypoplasia or dysplasia of lung Congenital pulmonary dysplasia Hypoplasia of lung Pulmonary hypoplasia Hypoplasia of lobe of lung Other specified dysplasia of lung Unspecified dysplasia of lung Excludes 1: pulmonary hypoplasia associated with short gestation (P28.0)	748.5
<u>Q33.8</u>	Other congenital malformations of lung (must specify) Aplasia of lung Bilobar right lung Congenital lobar emphysema Fusion of lobes of lung Lymphangiectasia of lungs Right lung with left lung bronchial pattern Trilobal left lung	748.69
<u>Q33.9</u>	Congenital malformations of lung, unspecified Congenital anomaly of lung	748.60
<u>Q34</u>	Other congenital malformations of respiratory system Excludes 2: congenital central alveolar hypoventilation syndrome (G47.35)	
<u>Q34.0</u>	Anomaly of pleura Congenital anomaly of pleural folds Abnormal communication between pericardial/pleural sacs	748.8
<u>Q34.1</u>	Congenital cyst of mediastinum	748.8
<u>Q34.8</u>	Other specified congenital malformation of respiratory system (must specify) Atresia of nasopharynx Immotile cilia syndrome Primary ciliary dyskinesia	748.8
<u>Q34.9</u>	Congenital malformation of respiratory system, unspecified (try not to use this code) Congenital absence of respiratory system Congenital anomaly of respiratory system, NOS	748.9

Congenital Malformations of the Cleft Lip and Cleft Palate (Q35-Q37)

Excludes 2: Robin's Syndrome (Q87.0)

ICD-10	Description	ICD-9
Q35	Cleft palate Excludes 1: cleft palate with cleft lip (Q37.-)	
Q35.1	Cleft hard palate Cleft hard palate, central Cleft hard palate, NOS Cleft hard palate, unilateral, left Cleft hard palate, unilateral, right Cleft hard palate, unilateral Cleft hard palate, bilateral	749.00
Q35.3	Cleft soft palate Cleft palate, submucous Submucous cleft palate Cleft soft palate alone, central Cleft soft palate alone, NOS Cleft soft palate alone, unilateral, left Cleft soft palate alone, unilateral, right Cleft soft palate alone, unilateral Cleft soft palate alone, bilateral	749.00
Q35.5	Cleft hard palate with cleft soft palate Complete cleft palate, unilateral, right Complete cleft palate, unilateral Complete cleft palate, bilateral Complete cleft palate, unilateral, left Complete cleft palate, central Uranostaphyloschisis	749.01 749.03
Q35.7	(m) Cleft uvula (m) Bifid uvula	749.02
Q35.9	Cleft palate, unspecified (try not to use this code) Cleft palate, NOS Cleft secondary palate Palatoschisis	749.00 749.02 749.04
Q36	Cleft lip Includes: cheiloschisis, congenital fissure of lip, harelip, labium leporinum Excludes 1: cleft lip with cleft palate (Q37.-)	
Q36.0	Cleft lip, bilateral Complete cleft lip, bilateral Incomplete cleft lip, bilateral	749.13 749.14
Q36.1	Cleft lip, median Central cleft lip	749.10
Q36.9	Cleft lip, unilateral Cleft lip, NOS Cleft gingiva Cleft gum Cleft lip, unilateral, left Cleft lip, unilateral, right Cleft lip, unspecified Complete cleft lip, unilateral Complete cleft lip, unilateral, left Complete cleft lip, unilateral, right Incomplete cleft lip, unilateral Incomplete cleft lip, unilateral, left Incomplete cleft lip, unilateral, right Incomplete form of cleft lip (microform)	749.10 749.11 749.12
Q37	Cleft palate with cleft lip	

	Includes: Cheilopatatoschisis	
<u>Q37.0</u>	Cleft hard palate with bilateral cleft lip <u>Cleft hard palate with cleft lip, NOS</u>	749.24 749.25
<u>Q37.1</u>	Cleft hard palate with unilateral cleft lip <u>Cleft hard palate with cleft lip, NOS</u>	749.22 749.25
<u>Q37.2</u>	Cleft soft palate with bilateral cleft lip	749.24 749.25
<u>Q37.3</u>	Cleft soft palate with unilateral cleft lip <u>Cleft soft palate with cleft lip, NOS</u>	749.22 749.25
<u>Q37.4</u>	Cleft hard and soft palate with bilateral cleft lip <u>Complete cleft palate and bilateral cleft lip</u>	749.23 749.24
<u>Q37.5</u>	Cleft hard and soft palate with unilateral cleft lip Cleft hard and soft palate with cleft lip, NOS <u>Complete cleft palate and cleft lip, unilateral, left</u> <u>Complete cleft palate and cleft lip, unilateral, right</u> <u>Complete cleft palate and cleft lip, unilateral</u> Complete cleft palate and cleft lip, NOS Uranostaphyloschisis with cleft lip	749.21 749.22
<u>Q37.8</u>	Unspecified cleft palate with bilateral cleft lip <u>Incomplete cleft palate and bilateral cleft lip</u> <u>Bilateral cleft lip, with any cleft palate</u>	749.24
<u>Q37.9</u>	Unspecified cleft palate with unilateral cleft lip Cleft palate with cleft lip, NOS <u>Cheilopatatoschisis</u> <u>Cleft palate with cleft lip</u> <u>Cleft palate with cleft lip, unspecified</u> <u>Other combinations cleft palate and lip</u>	749.20

Congenital Malformations of the Digestive System (Q38-Q45)

ICD-10	Descriptions	ICD-9
Q38	Other congenital malformations of tongue, mouth and pharynx Excludes 1: dentofacial anomalies (M26.-), macrostomia (Q18.4), microstomia (Q18.5)	
Q38.0	Congenital malformations of lips, NEC Van der Woude syndrome (m) <i>Congenital fistula of lip or pits</i> (m) <i>Congenital palsy of the lower lip</i> (m) <i>Long philtrum (only code if with Q86.-)</i> (m) <i>Lip tie</i> (m) <i>Notched lip (only code if with Q86.-)</i> (m) <i>Other lip anomaly (only code if with Q86.-)</i> (m) <i>Prominent philtrum (only code if with Q86.-)</i> Excludes 1: cleft lip (Q36.-), cleft lip with cleft palate (Q37.-), macrocheilia (Q18.6), microcheilia (Q18.7)	750.25
Q38.1	(m) <i>Ankyloglossia</i> (m) <i>Tongue tie</i> (m) <i>Short frenulum of tongue</i> (m) <i>Aberrant frenula</i>	750.0
Q38.2	(m) <i>Macroglossia (large tongue)</i> (m) <i>Congenital hypertrophy of tongue</i>	750.15
Q38.3	Other congenital malformations of tongue (must specify) Aglossia (absence of tongue) (m) <i>Bifid tongue</i> (m) <i>Congenital abnormal tongue position anomaly</i> (m) <i>Congenital adhesion of tongue</i> (m) <i>Congenital anomaly of tongue</i> (m) <i>Congenital fissure of tongue</i> (m) <i>Double tongue</i> (m) <i>Hypoglossia (small tongue)</i> (m) <i>Hypoplasia of tongue</i> (m) <i>Microglossia</i> (m) <i>Cleft tongue or split tongue</i> (m) <i>Dislocation or displacement of tongue</i> (m) <i>Glossoptosis</i>	750.10 750.11 750.12 750.13 750.16 750.19
Q38.4	Congenital malformations of salivary glands and ducts (must specify) Atresia of salivary glands and ducts Congenital absence of salivary glands and ducts Congenital accessory salivary glands and ducts Congenital fistula of salivary gland Congenital ranula Congenital salivary gland anomaly <i>Imperforate salivary duct</i> <i>Other anomaly of salivary glands or ducts</i>	750.21 750.22 750.23 750.24
Q38.5	Congenital malformations of palate, NEC (m) <i>Congenital absence of uvula</i> (m) <i>Congenital malformation of palate, NOS</i> (m) <i>Congenital high arched palate</i> Excludes 1: cleft palate (Q35.-), cleft palate with cleft lip (Q37.-)	750.26
Q38.6	(m) <i>Other congenital malformations of mouth (must specify)</i> (m) <i>Congenital malformations of mouth, NOS</i> (m) <i>Fordyce spots of mouth</i> (m) <i>Fordyce's disease</i> (m) <i>Low lying maxillary frenulum</i>	750.26
Q38.7	Congenital pharyngeal pouch Congenital diverticulum of pharynx	750.27

	Excludes 1: pharyngeal pouch syndrome (D82.1)	
Q38.8	Other congenital malformations of pharynx (must specify) Congenital malformation of pharynx, NOS Congenital anomaly of pharynx Congenital velopharyngeal dysfunction Congenital velopharyngeal impairment Congenital velopharyngeal incompetence Imperforate pharynx Do not report tonsil asymmetry	750.29
Q39	Congenital malformations of esophagus	
Q39.0	Atresia of esophagus without fistula Atresia of esophagus, NOS Esophageal atresia Imperforate esophagus	750.3
Q39.1	Atresia of esophagus with tracheoesophageal fistula Atresia of esophagus with broncho-esophageal fistula Esophageal atresia with tracheoesophageal fistula	750.3
Q39.2	Congenital trachea-esophageal fistula without atresia Congenital trachea-esophageal fistula, NOS Congenital trachea esophageal fistula without atresia of esophagus Esophagotracheal fistula, congenital T E fistula without esophageal atresia Tracheoesophageal fistula "h" type	750.3
Q39.3	Congenital stenosis and stricture of esophagus Congenital esophageal ring Congenital Schatzkis ring Congenital stenosis of esophagus	750.3
Q39.4	Esophageal web	750.3
Q39.5	Congenital dilatation of esophagus Congenital cardiospasm Congenital achalasia Giant esophagus	750.4
Q39.6	Congenital diverticulum of esophagus Congenital esophageal pouch	750.4
Q39.8	Other congenital malformations of esophagus (must specify) Congenital absence of esophagus Congenital displacement of esophagus Congenital duplication of esophagus Esophageal duplication Other tracheoesophageal anomaly	750.4
Q39.9	Congenital malformation of esophagus, unspecified	750.4
Q40	Other congenital malformations of upper alimentary tract	
Q40.0	Congenital hypertrophic pyloric stenosis Congenital or infantile constriction of pylorus Congenital or infantile hypertrophy of pylorus Congenital or infantile spasm of pylorus Congenital or infantile stenosis of pylorus Congenital or infantile stricture of pylorus Other congenital pyloric obstruction Do not report epileptic/infantile spasms	750.5
Q40.1	Congenital hiatus hernia Congenital hiatal hernia Congenital displacement of cardia through esophageal hiatus Excludes 1: congenital diaphragmatic hernia (Q79.0)	750.6
Q40.2	Other specified congenital malformations of stomach (must specify) Congenital displacement of stomach Congenital diverticulum of stomach	750.7

	Congenital hourglass stomach Congenital duplication of stomach Megalogastria Micogastria Displacement or transposition of stomach	
Q40.3	Congenital malformation of stomach, unspecified Congenital gastric anomaly	750.7
Q40.8	Other specified congenital malformations of upper alimentary tract	750.8
Q40.9	Congenital malformation of upper alimentary tract, unspecified Congenital anomaly of upper alimentary tract Congenital deformity of upper alimentary tract Congenital upper GI tract anomaly	750.9
Q41	Congenital absence, atresia or stenosis of small intestine Includes: congenital obstruction, occlusion or stricture of small intestine or intestine, NOS Excludes 1: cystic fibrosis with intestinal manifestation (E84.11), meconium ileus NOS (without cystic fibrosis) (P76.0)	
Q41.0	Congenital absence, atresia or stenosis of duodenum (must specify) Absence of duodenum/ duodenal absence Atresia of duodenum/ duodenal atresia Stenosis of duodenum/ duodenal stenosis	751.1
Q41.1	Congenital absence, atresia or stenosis of jejunum (must specify) Apple peel syndrome Imperforated jejunum Absence of jejunum Atresia of jejunum Stenosis of jejunum	751.1
Q41.2	Congenital absence, atresia or stenosis of ileum (must specify) Absence of ileum/ ileal absence Atresia of ileum/ ileal atresia Stenosis of ileum/ ileal stenosis	751.1
Q41.8	Congenital absence, atresia or stenosis of other specified parts of small intestine (must specify) Absence of other specified parts of small intestine (specify) Atresia of other specified parts of small intestine (specify) Stenosis of other specified parts of small intestine (specify)	751.1
Q41.9	Congenital absence, atresia and stenosis of small intestine, part unspecified Absence of small intestine Atresia of small intestine Stenosis of small intestine	751.1
Q42	Congenital absence, atresia and stenosis of large intestine Includes: congenital obstruction, occlusion and stricture of large intestine	
Q42.0	Congenital absence, atresia and stenosis of rectum with fistula (must specify) Absence of rectum with fistula Atresia of rectum with fistula Stenosis of rectum with fistula	751.2
Q42.1	Congenital absence, atresia and stenosis of rectum without fistula (must specify) Imperforated rectum Absence of rectum without fistula/rectal absence Atresia of rectum without fistula/ rectal atresia Stenosis of rectum without fistula/ rectal stenosis	751.2
Q42.2	Congenital absence, atresia and stenosis of anus with fistula (must specify) Absence of anus with fistula Atresia of anus with fistula Stenosis of anus with fistula Imperforated anus with fistula Congenital or infantile occlusion of anus with fistula Congenital infantile stricture of anus with fistula	751.2

<u>Q42.3</u>	Congenital absence, atresia and stenosis of anus without fistula (must specify) Imperforated anus <i>Absence of anus</i> <i>Atresia of anus</i> Congenital or infantile occlusion of anus without fistula Congenital infantile stricture of anus without fistula <i>Stenosis of anus</i>	751.2
<u>Q42.8</u>	Congenital absence, atresia and stenosis of other parts of large intestine (must specify) <i>Absence of appendix</i> <i>Atresia of appendix</i> <i>Stenosis of appendix</i>	751.2
<u>Q42.9</u>	Congenital absence, atresia, or stenosis of large intestine, part unspecified (try not to code) Absence of large intestine Atresia of colon Stenosis of colon	751.2
<u>Q43</u>	Other congenital malformations of intestine	
<u>Q43.0</u>	(m) Meckel's diverticulum (displaced) (hypertrophic) (m) Persistent vitelline duct (m) Persistent omphalomesenteric duct	751.0
<u>Q43.1</u>	Hirschsprung's disease Aganglionosis Congenital (aganglionic) megacolon Megacolon, congenital Long segment Hirschsprung's disease Short segment Hirschsprung's-Recto-Sigmoid	751.3
<u>Q43.2</u>	Other congenital functional disorders of colon (must specify) Congenital dilation of colon <i>Congenital megacolon, not aganglionic</i>	751.3
<u>Q43.3</u>	Congenital malformations of intestinal fixation (must specify) Congenital omentum, anomalous adhesions [bands] Congenital peritoneal adhesions [bands] Incomplete rotation of cecum and colon Insufficient rotation of cecum and colon Intestinal malrotation Jackson's membrane Malrotation of colon Malrotation of intestine Rotation failure of cecum and colon Universal mesentery <i>Anomalies of mesentery</i> <i>Ladd's bands</i> <i>Malrotation of cecum</i> <i>Malrotation of small intestine alone</i> <i>Other specified malrotation</i> <i>Unspecified malrotation</i>	751.4
<u>Q43.4</u>	Duplication of intestine Duplication of cyst ileum Duplication of anus Duplication of appendix Duplication of pylorus Duplication of cecum	751.5
<u>Q43.5</u>	Ectopic anus <i>Anterior anus</i>	751.5
<u>Q43.6</u>	Congenital fistula of rectum and anus	751.5

	Excludes 1: congenital fistula of anus with absence, atresia and stenosis (Q42.2), congenital fistula of rectum with absence, atresia and stenosis (Q42.0), congenital rectovaginal fistula (Q52.2), congenital urethrorectal fistula (Q64.73), pilonidal fistula or sinus (L05.-)	
Q43.7	Persistent cloaca Cloaca, NOS	751.5
Q43.8	Other specified congenital malformations of intestine (must specify) Congenital anal fissure Congenital blind loop syndrome Congenital diverticulitis, colon Congenital diverticulum, intestine Congenital rectal fissure Dolichocolon Megaloappendix Megaloduodenum Microcolon Transposition of appendix Transposition of colon Transposition of intestine Enterogenous cyst Duodenal web Volvus (m) Megaloappendix	751.5
Q43.9	Congenital malformation of intestine, unspecified	751.5
Q44	Congenital malformations of gallbladder, bile ducts and liver	
Q44.0	Agenesis, aplasia or hypoplasia of gallbladder Congenital absence of gallbladder Gallbladder hypoplasia Gallbladder aplasia Gallbladder agenesis	751.69
Q44.1	Other congenital malformations of gallbladder (must specify) Intrahepatic gallbladder Duplication gallbladder Floating gallbladder	751.69 751.60
Q44.2	Atresia of bile ducts Biliary atresia Carlois disease with obstruction Congenital dilatation of lobar intrahepatic bile duct with obstruction	751.61
Q44.3	Congenital stenosis or stricture of bile ducts Congenital biliary obstruction Congenital hypoplasia of bile duct (common) or passage	751.61
Q44.4	Choledochal cyst	751.69
Q44.5	Other congenital malformations of bile ducts (must specify) Accessory hepatic duct Biliary duct duplication Carlois disease without obstruction Congenital dilated bile duct Congenital dilatation of lobar intrahepatic bile duct Congenital malformation of bile duct, NOS Cystic duct duplication Agenesis or atresia of hepatic ducts Anomalies of biliary tract, NEC Other anomaly of hepatic or bile ducts	751.69 751.60
Q44.6	Cystic disease of liver Fibrocystic disease of liver	751.62

<u>Q44.7</u>	Other congenital malformation of liver (must specify) Accessory liver Alagille's syndrome Arteriohepatic dysplasia Congenital absence of liver Congenital focal nodular hyperplasia of liver Congenital malformation of liver, NOS Absence or agenesis of liver, partial Absence or agenesis of liver, total Floating liver Other anomaly of liver (m) <i>Congenital hepatomegaly</i>	751.60 751.69
<u>Q45</u>	Other congenital malformations of digestive system Excludes 2: congenital diaphragmatic hernia (Q79.0), congenital hiatus hernia (Q40.1)	
<u>Q45.0</u>	Agenesis, aplasia or hypoplasia of pancreas Congenital absence of pancreas Pancreatic agenesis Pancreatic aplasia Pancreatic hypoplasia	751.7
<u>Q45.1</u>	Annular pancreas	751.7
<u>Q45.2</u>	Congenital pancreatic cyst	751.7
<u>Q45.3</u>	Other congenital malformations of pancreas or pancreatic duct (must specify) Accessory pancreas Congenital malformation of pancreas or pancreatic duct, NOS Ectopic pancreas Ectopic pancreatic tissue Pancreatic heterotopia Excludes 1: congenital diabetes mellitus (E10.-), cystic fibrosis (E84.0-E84.9), fibrocystic disease of pancreas (E84.-), neonatal diabetes mellitus (P70.2)	751.7
<u>Q45.8</u>	Other specified congenital malformations of digestive system (must specify) Absence(complete) (partial) of alimentary tract, NOS Duplication of digestive system Malposition, congenital of digestive system Congenital malposition of digestive organs, NOS Cyst of mesenteric remnant Duplication of alimentary tract Ectopic digestive organs, NOS	751.8
<u>Q45.9</u>	Congenital malformation of digestive system, unspecified Congenital anomaly of digestive system Congenital deformity of digestive system	751.9

Congenital Malformations of Genital Organs (Q50-Q56)

ICD-10 codes 56.0 to 56.4 require chromosome results.

ICD- 10	Description	ICD-9
Q50	Congenital malformations of ovaries, fallopian tubes and broad ligaments	
Q50.0	Congenital absence of ovary [Only applicable to female patients.] Excludes 1: Turner's Syndrome (Q96.-)	
Q50.01	Congenital absence or agenesis of ovary, unilateral	752.0
Q50.02	Congenital absence or agenesis of ovary, bilateral	752.0
Q50.1	Developmental ovarian cyst Cystic ovary Multiple ovarian cysts	752.0
Q50.2	Congenital torsion of ovary	752.0
Q50.3	Other congenital malformations of ovary [Only applicable to female patients.]	
Q50.31	Accessory ovary	752.0
Q50.32	Ovarian streak 46.XX with streak gonads	752.0
Q50.39	Other congenital malformation of ovary Congenital malformation of ovary, NOS Ectopic ovary	752.0
Q50.4	Embryonic cyst of fallopian tube Fimbrial cyst	752.11
Q50.5	Embryonic cyst of broad ligament Cyst of hydatid of Morgagni Epoophoron cyst Parovarian cyst	752.11
Q50.6	Other congenital malformations of fallopian tube and broad ligament (must specify) Absence of fallopian tube and broad ligament Accessory fallopian tube and broad ligament Atresia of fallopian tube and broad ligament Congenital malformation of fallopian tube or board ligament, NOS	752.10 752.19
Q51	Congenital malformations of uterus and cervix [Only applicable to female patients.]	
Q51.0	Agenesis and aplasia of uterus Congenital absence of uterus	752.31
Q51.1	Doubling of uterus with doubling of cervix and vagina [Only applicable to female patients.]	
Q51.10	Doubling of uterus with doubling of cervix and vagina without obstruction Doubling of uterus with doubling of cervix and vagina, NOS	752.2
Q51.11	Doubling of uterus with doubling of cervix and vagina with obstruction	752.2
Q51.2	Other doubling of uterus [Only applicable to female patients.] Septate uterus	
Q51.21	Complete doubling of uterus Complete septate uterus	
Q51.22	Partial doubling of uterus Partial septate uterus	
Q51.28	Other and unspecified doubling of uterus Septate uterus, NOS	752.35
Q51.3	Bicornate uterus Bicornate uterus, complete or partial	752.34
Q51.4	Unicornate uterus Unicornate uterus with or without a separate uterine horn Uterus with only one functioning horn	752.33
Q51.5	Agenesis and aplasia of cervix Congenital absence of cervix	752.43
Q51.6	<i>(m) Embryonic cyst of cervix</i>	752.41
Q51.7	Congenital fistula between uterus and digestive and urinary tracts	752.49

	Congenital fistula between uterus and digestive tracts Congenital fistula between uterus and urinary tract Congenital uterointestinal fistula	752.39
<u>Q51.8</u>	Other congenital malformations of uterus and cervix [Only applicable to female patients.]	
<u>Q51.81</u>	Other congenital malformations of uterus [Only applicable to female patients.]	
<u>Q51.810</u>	Arcuate uterus Arcuatus uterus	752.36
<u>Q51.811</u>	Hypoplasia of uterus Infantile uterus	752.32
<u>Q51.818</u>	Other congenital malformations of uterus (must specify) Müllerian anomaly of uterus, NEC	752.39
<u>Q51.82</u>	Other congenital malformations of cervix [Only applicable to female patients.]	
<u>Q51.820</u>	Cervical duplication Congenital duplication of cervix	752.44
<u>Q51.821</u>	Hypoplasia of cervix	752.49
<u>Q51.828</u>	Other congenital malformations of cervix (must specify) Congenital imperforate cervix Congenital stenosis of cervical canal Congenital stenosis of uterine cervix <i>Congenital stenosis or stricture of cervical canal</i>	752.49
<u>Q51.9</u>	Congenital malformation of uterus and cervix, unspecified Congenital uterine anomaly Inadequate development of endometrium	752.39 752.49
<u>Q52</u>	Other congenital malformations of female genitalia [Only applicable to female patients.]	
<u>Q52.0</u>	Congenital absence of vagina Vaginal agenesis, total or partial	752.45
<u>Q52.1</u>	Doubling of vagina [Only applicable to female patients.] Excludes 1: doubling of vagina with doubling of uterus and cervix (<u>Q51.1-</u>)	
<u>Q52.10</u>	Doubling of vagina, unspecified Congenital duplication of vagina Congenital vaginal septum Septate vagina, NOS	752.47
<u>Q52.11</u>	Transverse vaginal septa Congenital transverse septated vagina	752.46
<u>Q52.12</u>	Longitudinal vaginal septum Congenital longitudinal vaginal septum Longitudinal septate vagina	
<u>Q52.120</u>	Longitudinal vaginal septum, non-obstructing (female only)	752.47
<u>Q52.121</u>	Longitudinal vaginal septum, obstructing, right side (female only)	752.47
<u>Q52.122</u>	Longitudinal vaginal septum, obstructing, left side (female only)	752.47
<u>Q52.123</u>	Longitudinal vaginal septum, microperforate, right side (female only)	752.47
<u>Q52.124</u>	Longitudinal vaginal septum, microperforate, left side (female only)	752.47
<u>Q52.129</u>	Other and unspecified longitudinal vaginal septum (female only)	752.47
<u>Q52.2</u>	Congenital rectovaginal fistula Excludes 1: cloaca (<u>Q43.7</u>)	752.49
<u>Q52.3</u>	(m) Imperforate hymen	752.42
<u>Q52.4</u>	Other congenital malformation of vagina (must specify) Congenital malformation of vagina, NOS (m) Canal of Nuck cyst, congenital (m) Congenital atresia of vagina (m) Embryonic vaginal cyst (m) Gartner's duct cyst (m) Vaginal adenosis (m) Cyst of vagina, vulva (m) Congenital stenosis or stricture of vagina	752.41 752.49
<u>Q52.5</u>	Fusion of labia	752.49

	(m) <i>Labial adhesions, congenital</i>	
<u>Q52.6</u>	Congenital malformation of clitoris (must specify) <i>Absence of clitoris</i> <i>Agenesis of clitoris</i> <i>(m) Clitoromegaly</i> <i>(m) Enlarged clitoris</i> <i>(m) Prominent clitoris</i>	752.49
<u>Q52.7</u>	Other and unspecified congenital malformations of vulva [Only applicable to female patients.]	
<u>Q52.70</u>	Unspecified congenital malformations of vulva Congenital anomaly of external female genitalia Congenital malformation of vulva, NOS <i>Fusion of vulva</i>	752.49
<u>Q52.71</u>	Congenital absence of vulva	752.49
<u>Q52.79</u>	Other congenital malformations of vulva <i>(m) Congenital cyst of vulva</i>	752.49
<u>Q52.8</u>	Other specified congenital malformation of female genitalia <i>(m) Vaginal or hymenal tags</i> <i>(m) Hypoplastic majora</i>	752.49
<u>Q52.9</u>	Congenital malformation of female genitalia, unspecified	752.9
<u>Q53</u>	Undescended and ectopic testicle [Only applicable to male patients.]	
<u>Q53.0</u>	Ectopic testis [Only applicable to male patients.]	
<u>Q53.00</u>	Ectopic testis, unspecified	752.51
<u>Q53.01</u>	Ectopic testis, unilateral <i>Ectopic testis, left only</i> <i>Ectopic testis, right only</i>	752.51
<u>Q53.02</u>	Ectopic testes, bilateral	752.51
<u>Q53.1</u>	Undescended testicle, unilateral [Only applicable to male patients.]	
<u>Q53.10</u>	Unspecified undescended testicle, unilateral (birth weight greater than 2,500 grams) <i>Cryptorchidism, left only</i> (birth weight greater than 2,500 grams) <i>Cryptorchidism, right only</i> (birth weight greater than 2,500 grams) <i>Cryptorchidism, unilateral</i> (birth weight greater than 2,500 grams) <i>Left undescended testicle</i> (birth weight greater than 2,500 grams) <i>Nonpalpable testis (specify laterality)</i> (birth weight greater than 2,500 grams) <i>Right undescended testicle</i> (birth weight greater than 2,500 grams)	752.51
<u>Q53.11</u>	Abdominal testis, unilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.111</u>	Intraabdominal testis, unilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.112</u>	Inguinal testis, unilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.12</u>	Ectopic perineal testis, unilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.13</u>	High scrotal testis, unilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.2</u>	Undescended testicle, bilateral [Only applicable to male patients.]	
<u>Q53.20</u>	Undescended testicle, unspecified, bilateral (birth weight greater than 2,500 grams) <i>Cryptorchidism, bilateral</i> (birth weight greater than 2,500 grams) <i>Nonpalpable testes</i> (birth weight greater than 2,500 grams) <i>Undescended testicle, bilateral</i> (birth weight greater than 2,500 grams)	752.51
<u>Q53.21</u>	Abdominal testis, bilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.211</u>	Intraabdominal testes, bilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.212</u>	Inguinal testes, bilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.22</u>	Ectopic perineal testis, bilateral (birth weight greater than 2,500 grams)	752.51
<u>Q53.23</u>	Bilateral high scrotal testes (birth weight greater than 2,500 grams)	
<u>Q53.9</u>	Undescended testicle, unspecified (birth weight greater than 2,500 grams) <i>Cryptorchidism, NOS</i> (birth weight greater than 2,500 grams) <i>Nonpalpable testis</i> (birth weight greater than 2,500 grams)	752.51
<u>Q54</u>	Hypospadias PLEASE SPECIFY DEGREE OR SITE OF HYPOSPADIAS [Only applicable to male patients.] Excludes 1: epispadias (Q64.0)	

<u>Q54.0</u>	Hypospadias, balanic Hypospadias, coronal Hypospadias, glandular Hypospadias 1 degree, glandular, coronal	752.61
<u>Q54.1</u>	Hypospadias, penile Hypospadias 2 degree, penile, subcoronal	752.61
<u>Q54.2</u>	Hypospadias, penoscrotal Hypospadias 3 degree, scrotal, penile scrotal	752.61
<u>Q54.3</u>	Hypospadias, perineal Hypospadias 3 degree, perineal, scrotal, penile scrotal	752.61
<u>Q54.4</u>	Congenital chordee (report only with hypospadias (ventral, dorsal, NOS)) <i>(m) Chordee without hypospadias</i>	752.63
<u>Q54.8</u>	Other hypospadias (must specify) Hypospadias with intersex state	752.61
<u>Q54.9</u>	Hypospadias, unspecified (must specify) Paraspadias	752.61
<u>Q55</u>	Other congenital malformations of male genital organs [Only applicable to male patients.] Excludes 1: congenital hydrocele (P83.5), hypospadias (Q54.-)	
<u>Q55.0</u>	Absence and aplasia of testis Congenital absence of testicle Monorchism	752.89
<u>Q55.1</u>	Hypoplasia of testis and scrotum Fusion of testes	752.89
<u>Q55.2</u>	Other and unspecified congenital malformations of testis and scrotum [Only applicable to male patients.]	
<u>Q55.20</u>	Unspecified congenital malformations of testis and scrotum Congenital malformation of testis or scrotum, NOS	752.9
<u>Q55.21</u>	Polyorchidism	752.89
<u>Q55.22</u>	<i>(m) Retractile testis</i>	752.52
<u>Q55.23</u>	Scrotal transposition Penoscrotal transposition	752.81
<u>Q55.29</u>	Other congenital malformations of testis and scrotum	752.89
<u>Q55.3</u>	Atresia of vas deferens Code first any associated cystic fibrosis (E84)	752.89
<u>Q55.4</u>	Other congenital malformations of vas deferens, epididymis, seminal vesicles and prostate (must specify) Absence or aplasia of prostate Absence or aplasia of spermatic cord Atresia of ejaculatory duct Appendix testis cyst Cysts of embryonic remnants Hydatid of morgagni cyst Wolfian duct cyst	752.89
<u>Q55.5</u>	Congenital absence and aplasia of penis	752.69
<u>Q55.6</u>	Other congenital malformations of penis	
<u>Q55.61</u>	Curvature of penis (lateral)	752.69
<u>Q55.62</u>	Hypoplasia of penis Micropenis	752.64
<u>Q55.63</u>	<i>(m) Congenital torsion of penis</i> Phimosis is not reportable Excludes 1: acquired torsion of penis (N48.82)	752.69
<u>Q55.64</u>	<i>(m) Buried penis</i> <i>(m) Concealed penis</i> <i>(m) Hidden penis</i> <i>(m) Webbed penis</i> Excludes 1: acquired buried penis (N48.83)	752.65

<u>Q55.69</u>	Other congenital malformation of penis Congenital malformation of penis, NOS Absent or hooded foreskin Incomplete foreskin	752.69
<u>Q55.7</u>	Congenital vasocutaneous fistula	752.89
<u>Q55.8</u>	Other specified congenital malformations of male genital organs	752.89
<u>Q55.9</u>	Congenital malformations of male genital organ, unspecified Congenital anomaly of male genital organ Congenital deformity of male genital organ	752.9
<u>Q56</u>	Indeterminate sex and pseudohermaphroditism PLEASE REPORT CHROMOSOME TEST RESULTS Excludes 1: 46,XX true hermaphrodite (Q99.1), androgen insensitivity syndrome (E34.5-), chimera 46,XXX/46,XY true hermaphrodite (Q99.0), female pseudohermaphroditism with adrenocortical disorder (E25.-), pseudohermaphroditism with specified chromosomal anomaly (Q96-Q99), pure gonadal dysgenesis (Q99.1)	
<u>Q56.0</u>	Hermaphroditism, NEC Ovotestis Gynandrom	752.7
<u>Q56.1</u>	Male pseudohermaphroditism, NEC 46, XY with streak gonads Male pseudohermaphroditism, NOS	752.7
<u>Q56.2</u>	Female pseudohermaphroditism, NEC Female pseudohermaphroditism, NOS	752.7
<u>Q56.3</u>	Pseudohermaphroditism, unspecified	752.7
<u>Q56.4</u>	Indeterminate sex, unspecified Ambiguous genitalia Disorder of sexual differentiation Sexual differentiation disorder	752.7

Congenital Malformations of the Urinary System (Q60-Q64)

ICD-10	Description	ICD-9
Q60	Renal agenesis and other reduction defects of kidney Includes: congenital absence of kidney, congenital atrophy of kidney, infantile atrophy of kidney	
Q60.0	Renal agenesis, unilateral Single kidney Solitary kidney	753.0
Q60.1	Renal agenesis, bilateral	753.0
Q60.2	Renal agenesis, unspecified	753.0
Q60.3	Renal hypoplasia, unilateral	753.0
Q60.4	Renal hypoplasia, bilateral	753.0
Q60.5	Renal hypoplasia, unspecified Oligomeganephronic hypoplasia of kidney	753.0
Q60.6	Potter's syndrome Oligohydramnios sequence	753.0
Q61	Cystic kidney disease Excludes 1: acquired cyst of kidney (N28.1), Potter's syndrome (Q60.6)	
Q61.0	Congenital renal cyst	
Q61.00	Congenital renal cyst, unspecified Cyst of kidney, NOS (congenital)	753.10
Q61.01	Congenital single renal cyst	753.11
Q61.02	Congenital multiple renal cysts	753.19
Q61.1	Polycystic kidney, infantile type Polycystic kidney, autosomal recessive	
Q61.11	Cystic dilation of collecting ducts	753.14
Q61.19	Other polycystic kidney, infantile type Autosomal recessive infantile polycystic kidney disease Polycystic kidney, autosomal recessive Congenital PKD	753.14
Q61.2	Polycystic kidney, adult type Polycystic kidney, autosomal dominant	753.13
Q61.3	Polycystic kidney, unspecified	753.12
Q61.4	Renal dysplasia Congenital renal dysplasia Multicystic dysplastic kidney Multicystic kidney (development) Multicystic kidney disease (MCDK) Multicystic renal dysplasia Dysplastic kidney, bilateral Renal dysplasia, bilateral Renal dysplasia, NOS; dysplastic kidney, NOS Unilateral dysplasia of kidney Excludes 1: polycystic kidney disease (Q61.11-Q61.3)	753.15 753.19
Q61.5	Medullary cystic kidney Medullary cystic kidney, juvenile type Medullary cystic kidney, adult type Medullary sponge kidney Nephronophthisis Sponge kidney, NOS	753.16 753.17
Q61.8	Other cystic kidney diseases (must specify) Fibrocystic kidney Fibrocystic renal degeneration of disease	753.19
Q61.9	Cystic kidney disease, unspecified (must specify) Cystic kidney disease Meckel-Gruber syndrome	753.10
Q62	Congenital obstructive defects of renal pelvis and congenital malformations of ureter	

<u>Q62.0</u>	Congenital hydronephrosis (swelling of kidney) Congenital hydronephrosis, bilateral Congenital hydronephrosis, left only Congenital hydronephrosis, NOS Congenital hydronephrosis, right only Congenital hydronephrosis, unilateral Pelvicaliectasis/Pelviectasis Pyelectasis	753.29
<u>Q62.1</u>	Congenital occlusion of ureter Atresia and stenosis of ureter	
<u>Q62.10</u>	Congenital occlusion of ureter, unspecified Stenosis of ureter Atresia, stricture, or stenosis of ureter, bilateral Atresia, stricture, or stenosis of ureter, left Atresia, stricture, or stenosis of ureter, NOS Atresia, stricture, or stenosis of ureter, right Atresia, stricture, or stenosis of ureter, unilateral Congenital occlusion of ureter, bilateral Congenital occlusion of ureter, left Congenital occlusion of ureter, right Congenital occlusion of ureter, unilateral Impervious ureter, bilateral Impervious ureter, left Impervious ureter, NOS Impervious ureter, right Impervious ureter, unilateral Ureteral valves, bilateral Ureteral valves, left Ureteral valves, NOS Ureteral valves, right Ureteral valves, unilateral	753.29
<u>Q62.11</u>	Congenital occlusion of ureteropelvic junction Stricture of pelviureteric junction Ureteropelvic junction (UPJ) obstruction/stenosis, left Ureteropelvic junction obstruction/stenosis, right Ureteropelvic junction obstruction/stenosis, unilateral Ureteropelvic junction obstruction/stenosis, bilateral	753.21
<u>Q62.12</u>	Congenital occlusion of ureterovesical orifice Ureterovesical junction obstruction/stenosis, NOS Ureterovesical junction obstruction/stenosis, left Ureterovesical junction obstruction/stenosis, right Ureterovesical junction obstruction/stenosis, unilateral Ureterovesical junction obstruction/stenosis, bilateral	753.22
<u>Q62.2</u>	Congenital megaureter Congenital dilation of ureter Congenital hydroureter, bilateral Congenital hydroureter, left Congenital hydroureter, NOS Congenital hydroureter, right Congenital hydroureter, unilateral Megaureter, NOS Megaureter, left Megaureter, right Megaureter, unilateral Megaureter, bilateral	753.22
<u>Q62.3</u>	Other obstructive defects of renal pelvis and ureter	
<u>Q62.31</u>	Congenital ureterocele, orthotopic	753.23

	Simple ureterocele Ureterocele is reportable; Hydrocele is NOT reportable	
Q62.32	Cecoureterocele Ectopic ureterocele	753.23
Q62.39	Other obstructive defects of renal pelvis ureter Congenital hydrourerter Congenital occlusion of ureter Congenital ureteral obstruction Congenital ureterovesical obstruction Duplicated collecting system with obstruction Ureteropelvic junction obstruction, NOS Urinary tract obstruction due to duplicated collecting system	753.20
Q62.4	Agenesis of ureter Congenital absence ureter	753.4
Q62.5	Duplication of ureter Accessory ureter Double ureter, duplex collecting system Duplication of collecting system	753.4
Q62.6	Malposition of ureter	
Q62.60	Malposition of ureter, unspecified	753.4
Q62.61	Deviation of ureter	753.4
Q62.62	Displacement of ureter	753.4
Q62.63	Anomalous implantation of ureter Ectopia of ureter Ectopic ureter Displaced ureteric orifice	753.4
Q62.69	Other malposition of ureter	753.4
Q62.7	Congenital vesico-uretero-renal reflux Ureteral reflux Variations of vesicoureteral reflux	753.4
Q62.8	Other congenital malformations of ureter (must specify) Anomaly of ureter, NOS Hypoplastic ureter, bilateral Hypoplastic ureter, left Hypoplastic ureter, NOS Hypoplastic ureter, right Other specified anomalies of ureter Unspecified anomaly of ureter Unspecified obstructive defects of renal pelvis and ureter	753.4
Q63	Other congenital malformations of kidney Excludes 1: congenital nephrotic syndrome (N04.-)	
Q63.0	Accessory kidney Double kidney and pelvis, pyelon duplex Trifid kidney (pelvis) Triple kidney and pelvis, pyelon triplex	753.3
Q63.1	Lobulated, fused or horseshoe kidney Fused kidney Horseshoe kidney Lobulated kidney	753.3
Q63.2	Ectopic kidney/ Pelvic Kidney Congenital displaced kidney Malrotation of kidney	753.3
Q63.3	Hyperplastic or giant kidney Compensatory hypertrophy of kidney	753.3
Q63.8	Other specified congenital malformations of kidney Congenital renal caluli/ calculus Congenital renal pelvis and ureteral defect Discoid kidney	753.3

	Duplicated collecting system without obstruction Inequal in size of kidney Megacalycosis, congenital	
Q63.9	Congenital malformation of kidney, unspecified	753.3
Q64	Other congenital malformations of urinary system	
Q64.0	Epispadias <i>Anaspadias</i> <i>Ventral meatus</i> Excludes 1: hypospadias (Q54.-) [Q64.0 is only applicable to male patients.]	752.62
Q64.10	Exstrophy of urinary bladder, unspecified Ectopia vesicae Exstrophy of bladder sequence	753.5
Q64.11	Supravesical fissure of urinary bladder	753.8
Q64.12	Cloacal exstrophy of urinary bladder Exstrophy of cloaca sequence	753.5
Q64.19	Other exstrophy of urinary bladder Estroversion of bladder	753.5
Q64.2	Congenital posterior urethral valves	753.6
Q64.3	Other atresia and stenosis of urethra and bladder neck	
Q64.31	Congenital bladder neck obstruction Congenital bladder neck stricture Congenital obstruction of vesicourethral orifice	753.6
Q64.32	Congenital stricture of urethra	753.6
Q64.33	Congenital stricture of urinary meatus	753.6
Q64.39	Other atresia and stenosis of urethra and bladder neck Atresia and stenosis of urethra and bladder neck, NOS	753.6
Q64.4	Malformation of urachus Cyst of urachus Fistula of urachus Patent urachus Prolapse of urachus	753.7
Q64.5	Congenital absence of bladder and urethra	753.8
Q64.6	Congenital diverticulum of bladder	753.8
Q64.7	Other and unspecified congenital malformations of bladder and urethra Excludes 1: congenital prolapse of bladder (mucosa) (Q79.4)	
Q64.70	Unspecified congenital malformations of bladder and urethra Malformation of bladder or urethra, NOS	753.9
Q64.71	Congenital prolapse of urethra	753.8
Q64.72	Congenital prolapse of urinary meatus	753.8
Q64.73	Congenital urethrorectal fistula	753.8
Q64.74	Double urethra	753.8
Q64.75	Double urinary meatus	753.8
Q64.79	Other congenital malformations of bladder and urethra	753.8
Q64.8	Other specified congenital malformations of urinary system	753.8
Q64.9	Congenital malformation of urinary system, unspecified	753.9

Congenital Malformations of the Musculoskeletal System (Q65-Q79)

ICD-10	Description	ICD 9
Q65	Congenital deformities of hip Excludes 1: clicking hip (R29.4)	
Q65.0	Congenital dislocation of hip, unilateral	
Q65.00	Congenital dislocation of unspecified hip, unilateral	754.30
Q65.01	Congenital dislocation of right hip, unilateral	754.30
Q65.02	Congenital dislocation of left hip, unilateral	754.30
Q65.1	Congenital dislocation of hip, bilateral	754.31

Q65.2	Congenital dislocation of hip, unspecified	754.30
Q65.3	Congenital partial dislocation of hip, unilateral	
Q65.30	Congenital partial dislocation of unspecified hip, unilateral Congenital subluxation of unspecified hip, unilateral	754.32
Q65.31	Congenital partial dislocation of right hip, unilateral Congenital subluxation of right hip	754.32
Q65.32	Congenital partial dislocation of left hip, unilateral Congenital subluxation of left hip	754.32
Q65.4	Congenital partial dislocation of hip, bilateral Congenital subluxation of hip, bilateral	754.33
Q65.5	Congenital partial dislocation of hip, unspecified Congenital subluxation of hip	754.32
Q65.6	Congenital unstable hip Congenital dislocatable hip Predislocation status of hip at birth, unilateral, left Predislocation status of hip at birth, unilateral, right Predislocation status of hip at birth, unilateral Preluxation of hip, congenital, left Preluxation of hip, congenital, right Preluxation of hip, congenital, unilateral Unstable hip, unilateral Unstable hip, unilateral, left Unstable hip, unilateral, right	754.32
Q65.8	Other congenital deformities of hip	
Q65.81	Congenital coxa valga	755.61
Q65.82	Congenital coxa vara	755.62
Q65.89	Other specified congenital deformities of hip (must specify) Anteversion of femoral neck/ femoral anteversion (do not report if normal x-ray) Congenital acetabular dysplasia Hip dysplasia, NOS Bilateral hip dysplasia Congenital flexion deformity, hip or thigh, left Congenital flexion deformity, hip or thigh, right Developmental dysplasia of hip (DDH) Unilateral hip dysplasia	755.63
Q65.9	Congenital deformity of hip, unspecified (try not to use; must specify)	755.63
Q66	Congenital deformities of feet Excludes 1: reduction defects of feet (Q72.-), valgus deformities (acquired) (M21.0-), varus deformities (acquired) (M21.1-)	
Q66.0	Congenital talipes equinovarus	
Q66.00	Congenital talipes equinovarus, unspecified foot Talipes equinovarus, NOS Talipes equinovarus, unilateral Talipes equinovarus, bilateral	754.51
Q66.01	Talipes equinovarus, right	754.51
Q66.02	Talipes equinovarus, left	754.51
Q66.1	Congenital talipes calcaneovarus	
Q66.10	Congenital talipes calcaneovarus, unspecified foot Talipes calcaneovarus, unilateral Talipes calcaneovarus, bilateral	754.59
Q66.11	Congenital talipes calcaneovarus, right	754.59
Q66.12	Congenital talipes calcaneovarus, left	754.59
Q66.2	Congenital metatarsus adductus Congenital metatarsus varus Congenital metatarsus primus varus	
Q66.21	Congenital metatarsus (primus) varus	

<u>Q66.211</u>	Congenital metatarsus primus varus, right foot	754.52
<u>Q66.212</u>	Congenital metatarsus primus varus, left foot	754.52
<u>Q66.219</u>	Congenital metatarsus primus varus, unspecified foot	754.52
<u>Q66.22</u>	Congenital metatarsus adductus Congenital metatarsus varus	
<u>Q66.221</u>	Congenital metatarsus adductus, right foot Congenital metatarsus varus, right foot	754.53
<u>Q66.222</u>	Congenital metatarsus adductus, left foot Congenital metatarsus varus, left foot	754.53
<u>Q66.229</u>	Congenital metatarsus adductus, unspecified foot Congenital metatarsus varus, unspecified foot	754.53
<u>Q66.3</u>	Other congenital varus deformities of feet Hallux varus, congenital Talipes varus <i>Cubitus varus deformity of feet</i>	
<u>Q66.30</u>	Other congenital varus deformities of feet, unspecified foot Talipes varus	754.59
<u>Q66.31</u>	Other congenital varus deformities of feet, right foot Talipes varus, right	754.59
<u>Q66.32</u>	Other congenital varus deformities of feet, left foot Talipes varus, left	754.59
<u>Q66.4</u>	Congenital talipes calcaneovalgus	
<u>Q66.40</u>	Congenital talipes calcaneovalgus, unspecified foot <i>Congenital talipes calcaneovalgus, unilateral</i> <i>Congenital talipes calcaneovalgus, bilateral</i>	754.62
<u>Q66.41</u>	Congenital talipes calcaneovalgus, right foot	754.62
<u>Q66.42</u>	Congenital talipes calcaneovalgus, left foot	754.62
<u>Q66.5</u>	Congenital pes planus Congenital flat foot Congenital rigid flat foot Congenital spastic (everted) flat foot <i>(m) Congenital rocker bottom foot</i> Excludes 1: pes planus, acquired (M21.4)	
<u>Q66.50</u>	Congenital pes planus, unspecified foot	754.61
<u>Q66.51</u>	Congenital pes planus, right foot	754.61
<u>Q66.52</u>	Congenital pes planus, left foot	754.61
<u>Q66.6</u>	Other congenital valgus deformities of feet (must specify) Congenital metatarsus valgus Hallus valgus Pes valgoplanus congenital Talipes valgus <i>Claw foot</i> <i>Talipes equinovalgus</i>	754.69
<u>Q66.7</u>	Congenital pes cavus <i>Talipes cavus</i>	
<u>Q66.70</u>	Congenital pes cavus, unspecified foot	754.71
<u>Q66.71</u>	Congenital pes cavus, right foot	754.71
<u>Q66.72</u>	Congenital pes cavus, left foot	754.71
<u>Q66.8</u>	Other congenital deformities of feet	
<u>Q66.80</u>	Congenital vertical talus deformity, unspecified foot	754.61
<u>Q66.81</u>	Congenital vertical talus deformity, right foot	754.61
<u>Q66.82</u>	Congenital vertical talus deformity, left foot	754.61

<u>Q66.89</u>	Other specified congenital deformities of feet (must specify) Brachydactyly of toes Brachymetatarsia Congenital asymmetric talipes Congenital contracture of toe Congenital talipes, NOS Congenital tarsal coalition Congenital clubfoot, NOS Fusion of calcaneus (heel bone) Tarsal coalitions (specify laterality) Anomalies of foot Astragalo-scaphoid synostosis Calcaneonavicular bar Clubfoot, NOS Congenital hallux valgus Congenital hallux varus Coalition of calcaneus Dorsiflexion of foot (fixed only) Other specified deformities of toes Plantar furrow Prominent heel Talipes calcaneus Talipes equinus Talonavicular synostosis (m) Hammer toe, congenital (m) Curly toe (m) Gap between toes (1-2) (m) Long toes (m) Overlapping toes (m) Recessed toes (4,5) (m) Short or broad great toe (m) Wide spaced first and second toes	
<u>Q66.9</u>	Congenital deformity of feet, unspecified (must specify) Congenital deformity of toe(s) (must specify)	
<u>Q66.90</u>	Congenital deformity of feet, unspecified, unspecified foot (must specify)	754.79
<u>Q66.91</u>	Congenital deformity of feet, unspecified, right foot (must specify)	754.79
<u>Q66.92</u>	Congenital deformity of feet, unspecified, left foot (must specify)	754.79
<u>Q67</u>	Congenital musculoskeletal deformities of head, face, spine and chest Excludes 1: congenital malformation syndromes classified to Q87.- , Potter's syndrome (Q60.6)	
<u>Q67.0</u>	(m) Congenital facial asymmetry (m) Asymmetry of face (m) Facial asymmetry (m) Head asymmetry	754.0
<u>Q67.1</u>	(m) Congenital compression facies	754.0
<u>Q67.2</u>	(m) Dolichocephaly (m) Long narrow head	754.0
<u>Q67.3</u>	(m) Plagiocephaly (m) Flat occiput	754.0
<u>Q67.4</u>	(m) Other congenital deformities of skull, face and jaw (must specify) (m) Congenital hemifacial atrophy or hypertrophy (m) Congenital depressions in skull (m) Deviation of nasal septum, congenital (m) Squashed or bent nose, congenital (m) Oxycephaly (m) Trigonocephaly (m) Metopic suture open to bregma (m) Large or small fontanelles	748.1 754.0

	(m) <i>Scaphocephaly (if due to craniosynostosis)</i> (m) <i>Third fontanel</i> (m) <i>Trigonocephaly, other head deformations without synostosis</i> Excludes 1: dentofacial anomalies [including malocclusion] (M26.-), syphilitic saddle nose (A50.5)	
Q67.5	Congenital deformity of spine Congenital postural scoliosis Congenital scoliosis, NOS Excludes 1: infantile idiopathic scoliosis (M41.0), scoliosis due to congenital bony malformation (Q76.3)	754.2
Q67.6	Pectus excavatum Congenital funnel chest	754.81
Q67.7	Pectus carinatum Congenital pigeon chest	754.82
Q67.8	Other congenital deformities of chest (must specify) Congenital deformity of chest wall, NOS Congenital anomaly of thoracic cage Shield chest (m) <i>Bell shaped chest</i>	754.89
Q68	Other congenital musculoskeletal deformities Excludes 1: reduction defects of limb(s) (Q71-Q73) Excludes 2: congenital myotonic chondrodystrophy (G71.13)	
Q68.0	Congenital deformity of sternocleidomastoid muscle Congenital anomaly of sternocleidomastoid muscle Congenital contracture of sternocleidomastoid (muscle) Congenital (sternomastoid) torticollis Sternomastoid tumor (congenital) Absent or hypoplastic sternocleidomastoid muscle Congenital wryneck Sternomastoid tumor	754.1
Q68.1	Congenital deformity of finger(s) and hand (must specify) Congenital club finger Congenital crooked finger Spade-like hand (congenital) <i>Anthrogryposis</i> <i>Flexion contractures of individual joints of the hand</i>	754.89
Q68.2	Congenital deformity of knee Congenital dislocation of knee Congenital genu recurvatum	755.64 754.41 754.40
Q68.3	Congenital bowing of femur Excludes 1: anteversion of femur (neck) (Q65.89)	754.42
Q68.4	Congenital bowing of tibia and fibula	754.43
Q68.5	Congenital bowing of long bones of leg, unspecified	754.44
Q68.6	Discoid meniscus Discoid meniscus of knee	717.5
Q68.8	Other specified congenital musculoskeletal deformities (must specify) Congenital deformity of clavicle Congenital deformity of elbow Congenital deformity of forearm Congenital deformity of scapula Congenital deformity of wrist Congenital dislocation of elbow Congenital dislocation of shoulder Congenital dislocation of wrist Congenital dislocation of radial head <i>Congenital elevation of scapula</i> <i>Generalized flexion contractures of lower limb joints</i>	756.9 755.51 755.52
Q69	Polydactyly	

<u>Q69.0</u>	Accessory finger(s) Radial polydactyly Accessory digits hand, NOS Polydactyly of fingers	755.01
<u>Q69.1</u>	Accessory thumb(s)	755.01
<u>Q69.2</u>	Accessory toe(s) (do not report if skin tag) Accessory hallux Polydactyly of toes Accessory big toe (preaxial) Accessory toes (postaxial) Accessory digits foot, NOS	755.02
<u>Q69.9</u>	Polydactyly, unspecified (must specify) Supernumerary digit(s), NOS Postaxial polydactyly, NOS Accessory digits, NOS (hand/foot not specified)	755.00
<u>Q70</u>	Syndactyly	
<u>Q70.0</u>	Fused fingers Complex syndactyly of fingers and/or thumb with synostosis	
<u>Q70.00</u>	Fused fingers and/or thumb , unspecified hand Syndactyly of fingers with fusion of bones	755.12
<u>Q70.01</u>	Fused fingers and/or thumb , right hand Syndactyly of fingers of right hand with fusion of bones	755.12
<u>Q70.02</u>	Fused fingers and/or thumb , left hand Syndactyly of fingers of left hand with fusion of bones	755.12
<u>Q70.03</u>	Fused fingers and/or thumb , bilateral Syndactyly of fingers, bilateral, with fusion of bones	755.12
<u>Q70.1</u>	(m) Webbed fingers (m) Simple syndactyly of fingers without synostosis	
<u>Q70.10</u>	(m) Webbed fingers, unspecified hand	755.11
<u>Q70.11</u>	(m) Webbed fingers, right hand	755.11
<u>Q70.12</u>	(m) Webbed fingers, left hand	755.11
<u>Q70.13</u>	(m) Webbed fingers, bilateral	755.11
<u>Q70.2</u>	Fused toes Complex syndactyly of toes with synostosis	
<u>Q70.20</u>	Fused toes, unspecified foot Syndactyly of toes with fusion of bones	755.14
<u>Q70.21</u>	Fused toes, right foot Syndactyly of toes of right foot, with fusion of bones	755.14
<u>Q70.22</u>	Fused toes, left foot Syndactyly of toes of left foot, with fusion of bones	755.14
<u>Q70.23</u>	Fused toes, bilateral Syndactyly of toes of both feet, with fusion of bones	755.14
<u>Q70.3</u>	(m) Webbed toes (m) Simple syndactyly of toes without synostosis	
<u>Q70.30</u>	(m) Webbed toes, unspecified foot (m) Simple syndactyly of toes	755.13
<u>Q70.31</u>	(m) Webbed toes, right foot	755.13
<u>Q70.32</u>	(m) Webbed toes, left foot	755.13
<u>Q70.33</u>	(m) Webbed toes, bilateral	755.13
<u>Q70.4</u>	Polysyndactyly, unspecified Excludes 1: specified syndactyly of hand and feet - code to specified conditions (<u>Q70.0-Q70.3</u>)	755.00 755.10
<u>Q70.9</u>	Syndactyly, unspecified Symphalangy, NOS Syndactyly of multiple and unspecified sites	755.10
<u>Q71</u>	Reduction defects of upper limb	

<u>Q71.0</u>	Congenital complete absence of upper limb Transverse deficiency of upper limb	
<u>Q71.00</u>	Congenital complete absence of unspecified upper limb Congenital complete absence of arm Transverse deficiency of arm Congenital amputation of upper limb, NOS Transverse hemimelia of upper limb	755.21
<u>Q71.01</u>	Congenital complete absence of right upper limb	755.21
<u>Q71.02</u>	Congenital complete absence of left upper limb	755.21
<u>Q71.03</u>	Congenital complete absence of upper limb, bilateral	755.21
<u>Q71.1</u>	Congenital absence of upper arm and forearm with hand present	
<u>Q71.10</u>	Congenital absence of unspecified upper arm and forearm with hand present Complete phocomelia of upper limb Proximal phocomelia of upper limb Distal phocomelia of upper limb (forearm missing, upper arm/hand present) Total absence of forearm only (radius and ulna)	755.23
<u>Q71.11</u>	Congenital absence of right upper arm and forearm with hand present	755.23
<u>Q71.12</u>	Congenital absence of left upper arm and forearm with hand present	755.23
<u>Q71.13</u>	Congenital absence of upper arm and forearm with hand present, bilateral Complete phocomelia of bilateral upper limbs	755.23
<u>Q71.2</u>	Congenital absence of both forearm and hand Transverse deficiency, radioulnar, complete or partial	
<u>Q71.20</u>	Congenital absence of both forearm and hand, unspecified upper limb	755.25
<u>Q71.21</u>	Congenital absence of both forearm and hand, right upper limb	755.25
<u>Q71.22</u>	Congenital absence of both forearm and hand, left upper limb	755.25
<u>Q71.23</u>	Congenital absence of both forearm and hand, bilateral	755.25
<u>Q71.3</u>	Congenital absence of hand and finger	
<u>Q71.30</u>	Congenital absence of unspecified hand and finger Congenital absence of hand Congenital deficiency of hand bones Congenital longitudinal deficiency of fingers Longitudinal deficiency of phalanges of hand Absence of finger(s), congenital (includes individual missing fingers/phalanges) Absence of preaxial fingers Absence of thumb	755.29
<u>Q71.31</u>	Congenital absence of right hand and finger Congenital longitudinal deficiency of right fingers Hypoplasia of right finger(s) Hypoplasia of right thumb	755.29
<u>Q71.32</u>	Congenital absence of left hand and finger Congenital longitudinal deficiency of left fingers Hypoplasia of left finger(s) Hypoplasia of left thumb	755.29
<u>Q71.33</u>	Congenital absence of hand and finger, bilateral Hypoplasia of fingers, bilateral Hypoplasia of thumbs, bilateral	755.29
<u>Q71.4</u>	Longitudinal reduction defect of radius (congenital) Radial clubhand	
<u>Q71.40</u>	Longitudinal reduction defect of unspecified radius Radial hemimelia Absence of radius Congenital absence of radius with or without absence of some distal elements Longitudinal deficiency, radial, complete or partial Preaxial (longitudinal) reduction deficiency of upper limb, radial ray defect	755.26
<u>Q71.41</u>	Longitudinal reduction defect of right radius	755.26
<u>Q71.42</u>	Longitudinal reduction defect of left radius	755.26

Q71.43	Longitudinal reduction of radius, bilateral	755.26
Q71.5	Longitudinal reduction defect of ulna	
Q71.50	Longitudinal reduction defect of unspecified ulna Longitudinal deficiency of ulna <i>Absence of ulna</i> Congenital absence ulna, with or without absence of some distal elements Longitudinal deficiency ulna, complete or partial Postaxial (longitudinal) reduction deficiency of upper limb, ulna ray defect	755.27
Q71.51	Longitudinal reduction defect of right ulna	755.27
Q71.52	Longitudinal reduction defect of left ulna	755.27
Q71.53	Longitudinal reduction defect of ulna, bilateral	755.27
Q71.6	Lobster-claw hand	
Q71.60	Lobster-claw hand, unspecified hand Cleft hand	755.58
Q71.61	Lobster-claw right hand Cleft hand, right hand	755.58
Q71.62	Lobster-claw left hand Cleft hand, left hand	755.58
Q71.63	Lobster-claw hand, bilateral Cleft hand, bilateral	755.58
Q71.8	Other reduction defect of upper limb	
Q71.81	Congenital shortening of upper limb	
Q71.811	Congenital shortening of right upper limb	755.20
Q71.812	Congenital shortening of left upper limb	755.20
Q71.813	Congenital shortening of upper limb, bilateral	755.20
Q71.819	Congenital shortening of unspecified upper limb	755.20
Q71.89	Other reduction defects of upper limb	
Q71.891	Other reduction defects of right upper limb Brachydactyly of right fingers	755.20
Q71.892	Other reduction defects of left upper limb Brachydactyly of left fingers	755.20
Q71.893	Other reduction defects of upper limb, bilateral	755.20
Q71.899	Other reduction defects of unspecified upper limb (must specify) Ectromelia NOS of upper limb Hemimelia of upper limb, NOS Longitudinal reduction deficiency of upper limb, unspecified ray defect	755.20
Q71.9	Unspecified reduction defect of upper limb	
Q71.90	Unspecified reduction defect of unspecified upper limb	755.20
Q71.91	Unspecified reduction defect of right upper limb Longitudinal deficiency of right upper arm	755.20
Q71.92	Unspecified reduction defect of left upper limb Longitudinal deficiency of left upper arm	755.20
Q71.93	Unspecified reduction defect of upper limb, bilateral	755.20
Q72	Reduction defects of lower limb	
Q72.0	Congenital complete absence of lower limb	
Q72.00	Congenital complete absence of unspecified lower limb Congenital complete absence of unspecified leg Transverse deficiency unspecified lower limb Transverse hemimelia of unspecified lower leg	755.31
Q72.01	Congenital complete absence of right lower limb Congenital complete absence of right leg Transverse deficiency right lower limb Transverse hemimelia of right leg	755.31
Q72.02	Congenital complete absence of left lower limb Congenital complete absence of left leg Transverse deficiency left lower limb	755.31

	Transverse hemimelia of left leg	
Q72.03	Congenital complete absence of lower limbs, bilateral Congenital complete absence of legs, bilateral Transverse deficiency lower limbs, bilateral Transverse hemimelia of lower legs, bilateral	755.31
Q72.1	Congenital absence of thigh and lower leg with foot present	
Q72.10	Congenital absence of unspecified thigh and lower leg with foot present Complete phocomelia of lower limb (upper and lower leg missing, foot present) Absence of lower leg only (femur and foot present) Distal phocomelia of lower limb (lower leg absent, femur/foot present) Proximal phocomelia of lower limb (femur absent, lower leg, foot present)	755.33
Q72.11	Congenital absence of right thigh and lower leg with foot present	755.33
Q72.12	Congenital absence of left thigh and lower leg with foot present	755.33
Q72.13	Congenital absence of thigh and lower leg with foot present, bilateral	755.33
Q72.2	Congenital absence of both lower leg and foot	
Q72.20	Congenital absence of both lower leg and foot, unspecified lower limb Transverse deficiency lower limb - knee level	755.35
Q72.21	Congenital absence of both lower leg and foot, right lower limb	755.35
Q72.22	Congenital absence of both lower leg and foot, left lower limb	755.35
Q72.23	Congenital absence of both lower leg and foot, bilateral	755.35
Q72.3	Congenital absence of foot and toe(s)	
Q72.30	Congenital absence of unspecified foot and toe(s) Longitudinal deficiency of phalanges of foot Longitudinal reduction defect of toes	755.38
Q72.31	Congenital absence of right foot and toe(s) Longitudinal deficiency of right toes Longitudinal reduction defect of right toes	755.38
Q72.32	Congenital absence of left foot and toe(s) Longitudinal deficiency of left toes Longitudinal reduction defect of left toes	755.38 755.39
Q72.33	Congenital absence of foot and toe(s), bilateral Longitudinal deficiency of bilateral toes Longitudinal reduction defect of bilateral toes	755.38 755.39
Q72.4	Longitudinal reduction defect of femur	
Q72.40	Longitudinal reduction defect of unspecified femur Longitudinal deficiency of unspecified femur Proximal focal deficiency of unspecified femur Absent unspecified femur, congenital	755.34
Q72.41	Longitudinal reduction defect of right femur Longitudinal deficiency of right femur Absent right femur, congenital	755.34
Q72.42	Longitudinal reduction defect of left femur Longitudinal deficiency of left femur Congenital absence of left femur	755.34
Q72.43	Longitudinal reduction defect of femur, bilateral Longitudinal deficiency of femur, bilateral Absent bilateral femurs, congenital	755.34
Q72.5	Longitudinal reduction defect of tibia	
Q72.50	Longitudinal reduction defect of unspecified tibia Congenital absence of tibia Longitudinal deficiency of tibia Tibial hemimelia	755.36
Q72.51	Longitudinal reduction defect of right tibia Longitudinal deficiency of right tibia Congenital absence of right tibia Tibial hemimelia, right	755.36

<u>Q72.52</u>	Longitudinal reduction defect of left tibia Longitudinal deficiency of left tibia Congenital absence of left tibia Tibial hemimelia, left	755.36
<u>Q72.53</u>	Longitudinal reduction defect of tibia, bilateral Congenital absence of tibia, bilateral Agenesis of tibia, bilateral Tibial hemimelia, bilateral	755.36
<u>Q72.6</u>	Longitudinal reduction defect of fibula Congenital absence of fibula	
<u>Q72.60</u>	Longitudinal reduction defect of unspecified fibula Congenital absence of fibula Fibular hemimelia Longitudinal deficiency of fibula Agenesis of unspecified fibula	755.37
<u>Q72.61</u>	Longitudinal reduction defect of right fibula Longitudinal deficiency of right fibula Congenital absence of right fibula Agenesis of right fibula	755.37
<u>Q72.62</u>	Longitudinal reduction defect of left fibula Longitudinal deficiency of left fibula Congenital absence of left fibula Agenesis of left fibula	755.37
<u>Q72.63</u>	Longitudinal reduction defect of fibula, bilateral Bilateral longitudinal deficiency of fibulas Congenital absence of fibula, bilateral Bilateral agenesis of fibula	755.37
<u>Q72.7</u>	Split foot Cleft foot Clawfoot or lobster claw foot	
<u>Q72.70</u>	Split foot, unspecified lower limb	755.67
<u>Q72.71</u>	Split foot, right lower limb Right split foot	755.67
<u>Q72.72</u>	Split foot, left lower limb Left split foot	755.67
<u>Q72.73</u>	Split foot, bilateral	755.67
<u>Q72.8</u>	Other reduction defects of lower limb	
<u>Q72.81</u>	Congenital shortening of lower limb	
<u>Q72.811</u>	Congenital shortening of right lower limb/leg	755.30
<u>Q72.812</u>	Congenital shortening of left lower limb/leg	755.30
<u>Q72.813</u>	Congenital shortening of lower limb/leg, bilateral	755.30
<u>Q72.819</u>	Congenital shortening of unspecified lower limb/leg	755.30
<u>Q72.89</u>	Other reduction defects of lower limb	
<u>Q72.891</u>	Other reductions defects of right lower limb Brachydactyly of right toes Longitudinal deficiency of right leg Longitudinal reduction defect of right leg	755.30
<u>Q72.892</u>	Other reductions defects of left lower limb Brachydactyly of left toes Longitudinal deficiency of left leg Longitudinal reduction defect of left leg	755.30
<u>Q72.893</u>	Other reduction defects of lower limb, bilateral	755.30
<u>Q72.899</u>	Other reduction defects of unspecified lower limb Longitudinal deficiency of lower limb Longitudinal reduction defect of leg	755.30
<u>Q72.9</u>	Unspecified reduction defect of lower limb	

<u>Q72.90</u>	Unspecified reduction defect of unspecified lower limb/leg	755.32
<u>Q72.91</u>	Unspecified reduction defect of right lower limb/leg	755.32
<u>Q72.92</u>	Unspecified reduction defect of left lower limb/leg	755.32
<u>Q72.93</u>	Unspecified reduction defect of lower limb/leg, bilateral	755.32
<u>Q73</u>	Reduction defects of unspecified limb (complete or partial)	
<u>Q73.0</u>	Congenital absence of unspecified limb(s)	755.4
<u>Q73.1</u>	Phocomelia, unspecified limb(s) Phocomelia, NOS	755.4
<u>Q73.8</u>	Other reduction defects of unspecified limb(s) (must specify) Ectromelia of limb, NOS Hemimelia of limb, NOS Longitudinal reduction deformity of unspecified limb(s) Reduction defect of limb, NOS Absent digits, NOS Amputation of unspecified limb Unspecified reduction defect of unspecified limb	755.4
<u>Q74</u>	Other congenital malformations of limb(s) Excludes 1: polydactyly (Q69.-), reduction defect of limb (Q71-Q73), syndactyly (Q70.-)	
<u>Q74.0</u>	Other congenital malformations of upper limb(s), including shoulder girdle (must specify) Accessory carpal bones Cleidocranial dysostosis Congenital abnormal fusion of carpal bone Congenital carpal coalition Congenital elevation of scapula Congenital glenoid dysplasia Congenital hypoplasia of ulna Congenital pseudarthrosis of clavicle Congenital trigger finger and trigger thumb Madelung's deformity Radioulnar synostosis Sprengel's deformity Triphalangeal thumb Anomalies of hand (must specify) Anomalies of elbow and upper arm (must specify) Anomalies of shoulder (must specify) Anomalies of wrist (must specify) Bifid thumb Cubitus valgus, congenital Cubitus varus, congenital Hyperextensibility of upper limb Hypoplasia of arms Hypoplasia of upper limb Other specified deformity of hands Radioulnar dysostosis <i>(m) Camptodactyly</i> <i>(m) Clinodactyly</i> <i>(m) Macrodactyly of finger(s)</i> <i>(m) Anomalies of finger</i> <i>(m) Brachydactyly</i> <i>(m) Incurving fingers</i> <i>(m) Long fingers</i> <i>(m) Long fingers and toes</i> <i>(m) Overlapping fingers</i> <i>(m) Tapered fingers</i>	755.53 755.54 755.56 755.57 755.59
<u>Q74.1</u>	Congenital malformation of knee (and joint) (must specify) Bifid patella Bipartite patella (kneecap)	755.64

	<p>Congenital absence of patella Congenital dislocation of patella (knee-cap) Congenital genu valgum (knock-knee) Congenital genu varum (bowleg) Rudimentary patella Excludes 1: congenital dislocation of knee (Q68.2), congenital genu recurvatum (Q68.2), nail patella syndrome (Q87.2)</p>	
Q74.2	<p>Other congenital malformations of lower limb(s), including pelvic girdle (must specify) Accessory tarsal bones Accessory tarsal navicular bone Congenital fusion of sacroiliac joint Congenital malformation of ankle joint Congenital malformation of sacroiliac joint Anomalies of ankle (must specify) Anomalies of pelvis (must specify) Anomalies of upper leg (must specify) Congenital deformity of sacroiliac (joint) Deformity of leg, NOS Dorsiflexion of foot Femoral hypoplasia syndrome Hypoplasia of feet Other specified anomaly of lower limb (must specify) <i>(m) Congenital internal tibial torsion</i> <i>(m) Macrodactylia of toe(s)</i> <i>(m) Anomalies of lower leg</i> <i>(m) Congenital angulation of tibia</i> Excludes 1: anteverision of femur (neck) (Q65.89)</p>	755.69
Q74.3	Arthrogryposis multiplex congenita	754.89
Q74.8	<p>Other specified congenital malformations of limb(s) (must specify) Larsen's syndrome</p>	755.8
Q74.9	<p>Unspecified congenital malformations of limb(s) (try not to use; must specify) Congenital anomaly of limb(s), NOS Dysplasia of limb Anomalies of forearm, NOS Other anomaly of whole arm Unspecified anomalies of upper limb</p>	755.9
Q75	<p>Other congenital malformations of skull and face bones Excludes 1: congenital malformation of face NOS (Q18.-), congenital malformation syndromes classified to (Q87.-), dentofacial anomalies [including malocclusion] (M26.-), musculoskeletal deformities of head and face (Q67.0-Q67.4), skull defects associated with congenital anomalies of brain such as: anencephaly (Q00.0), encephalocele (Q01.-), and hydrocephalus (Q03.-), microcephaly (Q02)</p>	
Q75.0	<p>Craniosynostosis Acrocephaly Complex craniosynostosis Craniosynostosis, NOS Imperfect fusion of skull Simple craniosynostosis Basilar craniosynostosis Closed skull sutures, NOS Cloverleaf skull (Keeblatt) Coronal craniosynostosis Kleeblattschädel Lambdoidal Metopic craniosynostosis Premature closure of cranial sutures Sagittal craniosynostosis</p>	756.0

Q75.1	Craniofacial dysostosis Crouzon's disease Crouzon syndrome	756.0
Q75.2	Hypertelorism Orbital hypertelorism Wide set eyes	756.0
Q75.3	Macrocephaly Macroencephaly	756.0
Q75.4	Mandibulofacial dysostosis Franceschetti syndrome Treacher Collins syndrome	756.0
Q75.5	Oculomandibular Dysostosis	756.0
Q75.8	Other specified congenital malformations of skull and face bones (must specify) Asymmetry of head (skull bones) Absence of skull bone, congenital Absence, cranial bone Basilar skull invagination Congenital deformity of forehead Delayed closure of anterior fontanel Forehead deformity, congenital Invagination of basilar skull Late closure of anterior fontanel Oculoauricular vertebral dysplasia Platybasia Cleft mandible malar hypoplasia Goldenhar syndrome Hallermann-Streiff syndrome Hemifacial microsomia Oculoauriculovertebral (OAV) dysplasia Oculomandibulofacial syndrome Orbital dystopia Other craniofacial syndromes Prominent occiput (m) Bony occipital spur (m) Frontal bossing (m) Hypotelorism (m) Localized skull defects (m) Midfacial hypoplasia	756.0
Q75.9	Congenital malformation of skull and face bones, unspecified (try not to use; must specify) Congenital anomaly of face bones, NOS Congenital anomaly of skull, NOS	756.0
Q76	Congenital malformations of spine and bony thorax Excludes 1: congenital musculoskeletal deformities of spine and chest (Q67.5 - Q67.8)	
Q76.0	Spina bifida occulta Excludes 1: meningocele (spinal) (Q05.-), spina bifida (aperta) (cystica) (Q05.-)	756.17
Q76.1	Klippel-Feil syndrome Cervical fusion syndrome Klippel feil sequence Thoracolumbar kissing spine Wildervanck syndrome Cervicooculaoaccoustic syndrome	756.16
Q76.2	Congenital spondylolisthesis Congenital spondylolysis Congenital cervical spondylolisthesis Congenital lumbar spondylolisthesis Congenital lumbosacral spondylolysis	756.12

	Prespondylolisthesis, lumbosacral Spondylosis, lumbosacral Excludes 1: spondylolisthesis (acquired) (M43.1-), spondylosis (acquired) (M43.0-)	
Q76.3	Congenital scoliosis due to congenital bony malformation Hemivertebra fusion or failure of segmentation with scoliosis	754.2
Q76.4	Other congenital malformations of spine, not associated with scoliosis	
Q76.41	Congenital kyphosis	
Q76.411	Congenital kyphosis, occipito-atlanto-axial region	756.19
Q76.412	Congenital kyphosis, cervical region	756.19
Q76.413	Congenital kyphosis, cervicothoracic region	756.19
Q76.414	Congenital kyphosis, thoracic region	756.19
Q76.415	Congenital kyphosis, thoracolumbar region	756.19
Q76.419	Congenital kyphosis, unspecified region	756.19
Q76.42	Congenital lordosis	
Q76.425	Congenital lordosis, thoracolumbar region	754.2
Q76.426	Congenital lordosis, lumbar region	754.2
Q76.427	Congenital lordosis, lumbosacral region	754.2
Q76.428	Congenital lordosis, sacral and sacrococcygeal region	754.2
Q76.429	Congenital lordosis, unspecified region	754.2
Q76.49	Other congenital malformations of spine, not associated with scoliosis (must specify) Agenesis of sacrum Aplasia odontoid Atlanto-occipital fusion Caudal agenesis (incomplete development of lower spine) Congenital absence of vertebra, NOS Congenital fusion of spine, NOS Congenital malformation of lumbosacral (joint) (region), NOS Congenital malformation of spine, NOS Dysgenesis, odontoid Dysraphism, congenital Dysraphism, cervical (neck) spine Fusion of atlantooccipital spine Hemivertebra, NOS Malformation of spine, NOS Occipitoatlantoaxial spine ankylosis Platyspondylisis, NOS Sacral agenesis Supernumerary vertebra, NOS Agenesis of cervical vertebrae Agenesis of lumbar vertebrae Agenesis of thoracic vertebrae Hemivertebrae, cervical vertebrae Hemivertebrae, thoracic vertebrae Hemivertebrae, lumbar vertebrae Sacral mass, NOS Sacrococcygeal anomaly	756.19 756.13 756.15 756.14
Q76.5	(m) Cervical rib (m) Supernumerary rib in cervical region	756.2
Q76.6	Other congenital malformations of ribs (must specify) Accessory rib Congenital absence of rib Congenital fusion of ribs Congenital malformation of ribs, NOS Excludes 1: short rib syndrome (Q77.2)	756.3
Q76.7	Congenital malformations of sternum (must specify) Congenital absence of sternum Sternum bifidum	756.3

	Anomalies of thoracic cage, unspecified Congenital fissure of sternum Double ossification center in the manubrium Short sternum	
Q76.8	Other congenital malformations of bony thorax	756.3
Q76.9	Congenital malformation of bony thorax, unspecified	756.3
Q77	Osteochondrodysplasia with defects of growth of tubular bones and spine Excludes 1: mucopolysaccharidosis (E76.0-E76.3) Excludes 2: congenital myotonic chondrodytrophy (G71.13)	
Q77.0	Achondrogenesis Hypochondrogenesis	756.4
Q77.1	Thanatophoric short stature Thanatophoric dysplasia	756.4
Q77.2	Short rib syndrome Asphyxiating thoracic dysplasia (Jeune) Short rib dysplasia	756.3
Q77.3	Chondrodysplasia punctata Chondrodysplasia punctata (stippled epiphyses) group Conradi syndrome Excludes 1: Rhizomelic chondrodysplasia punctata (E71.43)	756.59
Q77.4	Achondroplasia Hypochondroplasia Osteosclerosis congenital	756.4
Q77.5	Diastrophic dysplasia	756.4
Q77.6	Chondroectodermal dysplasia Ellis-van Creveld syndrome	756.55
Q77.7	Spondyloepiphyseal dysplasia Spondyloepimetaphyseal disorder	756.4
Q77.8	Other osteochondrodysplasia with defects of growth of tubular bones and spine	756.4
Q77.9	Osteochondrodysplasia with defects of growth of tubular bones and spine, unspecified	756.4
Q78	Other osteochondrodysplasias Excludes 2: congenital myotonic chondrodytrophy (G71.13)	
Q78.0	Osteogenesis imperfecta Brittle bones disease Fragilitas ossium Osteopsathyrosis	756.51
Q78.1	Polyostotic fibrous dysplasia Albright (McCune) (Sternberg) syndrome Osteitis fibrosa disseminata	756.54
Q78.2	Osteopetrosis Albers-Schönberg syndrome Osteosclerosis, NOS Marble bones	756.52
Q78.3	Progressive diaphyseal dysplasia (multiple epiphyseal dysplasia) Camurati-Engelmann syndrome Engelmann syndrome	756.56
Q78.4	Enchondromatosis Maffucci's syndrome Multiple enchondromatosis Ollier's disease	756.4
Q78.5	Metaphyseal dysplasia Pyle metaphyseal dysplasia Pyle's syndrome	756.59
Q78.6	Multiple congenital exostoses Diaphyseal aclasis	756.6
Q78.8	Other specified osteochondrodysplasia Dysplasia of epiphysis	756.59

	Epiphyseal dysplasia Homozygous Leri Weill dyschondrosteosis syndrome Langer mesomelic dysplasia syndrome Osteopoikilosis Melnick-Needles syndrome	
Q78.9	Osteochondrodysplasia, unspecified Chondrodysplasia Chondrodystrophy, NOS Osteodystrophy, NOS	756.50
Q79	Congenital malformations of musculoskeletal system, NEC Excludes 2: congenital (sternomastoid) torticollis (Q68.0)	
Q79.0	Congenital diaphragmatic hernia Excludes 1: congenital hiatus hernia (Q40.1)	756.6
Q79.1	Other congenital malformations of diaphragm (must specify) Absence of diaphragm Congenital malformation of diaphragm, NOS Eventration of diaphragm	756.6
Q79.2	Exomphalos Omphalocele Excludes 1: umbilical hernia (K42.-)	756.72
Q79.3	Gastroschisis	756.73
Q79.4	Prune belly syndrome Congenital prolapse of bladder mucosa Eagle-Barrett syndrome Early urethral obstruction sequence	756.71
Q79.5	Other congenital malformations of abdominal wall Excludes 1: umbilical hernia (K42.-)	
Q79.51	Congenital hernia of bladder	756.71
Q79.59	Other congenital malformations of abdominal wall (must specify) (<i>m</i>) Diastasis recti (<i>m</i>) Unspecified anomaly of abdominal wall	756.79
Q79.6	Ehlers-Danlos syndromes Cutis laxa hyperelastica	
Q79.60	Ehlers-Danlos syndrome, unspecified	756.83
Q79.61	Classical Ehlers-Danlos syndrome Classical EDS (cEDS)	756.83
Q79.62	Hypermobile Ehlers-Danlos syndrome Hypermobile EDS (hEDS)	756.83
Q79.63	Vascular Ehlers-Danlos syndrome Vascular EDS (vEDS)	756.83
Q79.69	Other Ehlers-Danlos syndromes (must specify)	756.83
Q79.8	Other congenital malformations of musculoskeletal system (must specify) Absence of muscle Absence of pectoral muscle, congenital Absence of tendon Accessory muscle Amyoplasia, disruptive sequence Amyotrophia congenital Congenital constricting bands Congenital shortening of tendon Contracture of gastrocnemius, congenital Hereditary osteoonychodysplasia Nail-patella syndrome Poland anomalad Poland syndrome Popliteal pterygium syndrome Schwartz Jampel syndrome Waardenburg syndrome	756.9 756.81 756.82

	Hypoplastic Depressor Angularis Muscle (face) Other absent or hypoplastic muscle Other specified anomaly muscle, tendon, fascia, connective tissue	
<u>Q79.9</u>	Congenital malformations of musculoskeletal system, unspecified (try not to use; must specify) Unspecified anomaly of bone Unspecified anomaly of cartilage Unspecified anomaly of connective tissue Unspecified anomaly of muscle Unspecified anomaly of musculoskeletal system Unspecified anomaly of tendon	756.9

Other Congenital Malformations (Q80-Q89)

ICD-10	Description	ICD-9
Q80	Congenital ichthyosis Excludes 1: Refsum's disease (G60.1)	
Q80.0	Ichthyosis vulgaris	757.1
Q80.1	X-linked ichthyosis Placental sulfatase deficiency (X-linked steryl-sulfatase deficiency) in a female	757.1
Q80.2	Lamellar ichthyosis Collodion baby	757.1
Q80.3	Congenital bullous ichthyosiform erythroderma Ichthyosiform erythroderma	757.1
Q80.4	Harlequin fetus	757.1
Q80.8	Other congenital ichthyosis	757.1
Q80.9	Congenital ichthyosis, unspecified Keratitis ichthyosis and deafness (KID) syndrome Netherton syndrome	757.1
Q81	Epidermolysis bullosa	
Q81.0	Epidermolysis bullosa simplex Excludes 1: Cockayne's syndrome (Q87.19)	757.39
Q81.1	Epidermolysis bullosa letalis Herlitz' syndrome	757.39
Q81.2	Epidermolysis bullosa dystrophica	757.39
Q81.8	Other epidermolysis bullosa	757.39
Q81.9	Epidermolysis bullosa, unspecified	757.39
Q82	Other congenital malformations of skin Excludes 1: acrodermatitis enteropathica (E83.2), congenital erythropoietic porphyria (E80.0), pilonidal cyst or sinus (L05.-), Sturge-Weber (-Dimitri) syndrome (Q85.8)	
Q82.0	Hereditary lymphedema Hereditary edema of leg(s) Meige syndrome Primary congenital lymphedema Hereditary trophefema Milroy's disease	757.0
Q82.1	Xeroderma pigmentosum	757.33
Q82.2	Congenital cutaneous mastocytosis Congenital diffuse cutaneous mastocytosis Congenital maculopapular cutaneous mastocytosis Congenital urticaria pigmentosa Urticaria pigmentosa Excludes 1: cutaneous mastocytosis NOS (D47.01), diffuse cutaneous mastocytosis (with onset after newborn period) (D47.01), malignant mastocytosis (C96.2-), systemic mastocytosis (D47.02), urticaria pigmentosa (non-congenital) (with onset after newborn period) (D47.01)	757.33
Q82.3	Incontinentia pigmenti	757.33
Q82.4	Ectodermal dysplasia (anhidrotic) Hay Wells syndrome of ectodermal dysplasia X-linked type ectodermal dysplasia Other specified ectodermal dysplasias Excludes 1: Ellis-van Creveld syndrome (Q77.6)	757.31
Q82.5	(m) Congenital non-neoplastic nevus (m) Birthmark, NOS (m) Flammeus nevus (m) Nevus anemicus (m) Portwine nevus (m) Sanguineous nevus (m) Strawberry nevus (m) Vascular nevus, NOS	757.32

	(m) <i>Verrucous nevus</i> (m) <i>Angel bite or kiss</i> (m) <i>Salmon patch</i> (m) <i>Stork bites</i> (m) <i>Vascular hamartomas</i> Excludes 2: Café au lait spots (L81.3), lentigo (L81.4), nevus NOS (D22.-), araneus nevus (I78.1), melanocytic nevus (D22.-), pigmented nevus (D22.-), spider nevus (I78.1), stellar nevus (I78.1)	
Q82.6	(m) <i>Congenital sacral dimple</i> (m) <i>Parasacral dimple</i> Excludes 2: pilonidal cyst with abscess (L05.01), pilonidal cyst without abscess (L05.91)	757.39
Q82.8	Other specified congenital malformations of skin (must specify) Acrokeratosis verruciformis Benign familial pemphigus [Hailey-Hailey disease] Bloom syndrome Congenital poikiloderma Cutis laxa (hyperelastica) Goltz syndrome Goltz-Gorlin (dermal hypoplasia) syndrome Howell Evans syndrome Inherited keratosis palmaris et plantaris Keratosis follicularis [Darier-White] <i>Absence of skin</i> <i>Cutis aplasia</i> (m) <i>Abnormal palmar creases</i> (m) <i>Darier disease</i> (m) <i>Mongolian spot</i> (m) <i>Porokeratosis</i> (m) <i>Cutis marmorata</i> (m) <i>Cutis marmorata telangiectatica congenital (CMTC)</i> (m) <i>Depigmented or hyperpigmented spot</i> (m) <i>Extra or absent creases</i> (m) <i>Hyperpigmented areas</i> (m) <i>Sidney line</i> (m) <i>Simian creases, transverse palmar creases</i> (m) <i>Single transverse palmar crease</i> <i>Congenital scars and non-ear skin tags are not reportable</i> Excludes 1: Ehlers-Danlos syndromes (Q79.6-)	757.39
Q82.9	Congenital malformation of skin, unspecified	757.39
Q83	Congenital malformations of breast Excludes 2: absence of pectoral muscle (Q79.8), hypoplasia of breast (N64.82), micromastia (N64.82)	
Q83.0	Congenital absence of breast with absent nipple	757.6
Q83.1	(m) <i>Accessory breast</i> (m) <i>Supernumerary breast</i>	757.6
Q83.2	Absent nipple	757.6
Q83.3	(m) <i>Accessory nipple (congenital)</i> (m) <i>Supernumerary nipple</i>	757.6
Q83.8	Other congenital malformations of breast (must specify) Congenital absence of breast <i>Hypoplastic breast with hypoplastic nipple</i> (m) <i>Asymmetry of breasts, congenital</i> (m) <i>Widely spaced nipples</i> (m) <i>Small nipple (hypoplastic)</i>	757.6
Q83.9	Congenital malformation of breast, unspecified (try not to use)	757.6
Q84	Other congenital malformations of integument	

<u>Q84.0</u>	(m) Congenital alopecia (m) Congenital atrichosis	757.4
<u>Q84.1</u>	(m) Congenital morphological disturbances of hair, NEC (m) Beaded hair (m) Monilethrix (m) Pili annulati (m) Pili torti (m) Twisted hair (m) Taenzer's hair Excludes 1: Menkes' kinky hair syndrome (E83.0)	757.4
<u>Q84.2</u>	(m) Other congenital malformations of hair (must specify) (m) Congenital hypertrichosis (m) Congenital malformation of hair, NOS (m) Persistent lanugo (m) Hirsutism (m) Aberrant scalp hair patterning (m) Low hair line (m) Other specified anomaly of hair	757.4
<u>Q84.3</u>	(m) Anonychia (m) Absent nails Excludes 1: nail patella syndrome (Q87.2)	757.5
<u>Q84.4</u>	(m) Congenital leukonychia	757.5
<u>Q84.5</u>	(m) Enlarged and hypertrophic nails (m) Congenital onychauxis (m) Pachyonychia (m) Hyperconvex or thickened toenails	757.5
<u>Q84.6</u>	Hypoplastic, hyperconvex, or duplicated fingernails (m) Other congenital malformations of nails (must specify) (m) Congenital clubnail (m) Congenital koilonychias (m) Congenital malformations of nail, NOS (m) Hypoplastic (small) fingernails and/or toenails	757.5
<u>Q84.8</u>	Other specified congenital malformations of integument (must specify) Aplasia cutis congenital (m) Scalp defects	757.8
<u>Q84.9</u>	(m) Congenital malformation of integument, unspecified (m) Congenital anomaly of integument, NOS (m) Congenital deformity of integument, NOS (m) Unspecified anomaly of hair (m) Unspecified anomaly of nail	757.9
<u>Q85</u>	Phakomatoses, NEC Excludes 1: ataxia telangiectasia [Louis-Bar] (G11.3), familial dysautonomia [Riley-Day] (G90.1)	
<u>Q85.0</u>	Neurofibromatosis (nonmalignant)	
<u>Q85.00</u>	Neurofibromatosis, unspecified	237.70
<u>Q85.01</u>	Neurofibromatosis, Type 1 Von Recklinghausen disease	237.71
<u>Q85.02</u>	Neurofibromatosis, Type 2 Acoustic neurofibromatosis	237.72
<u>Q85.03</u>	Schwannomatosis	237.73
<u>Q85.09</u>	Other neurofibromatosis	237.79
<u>Q85.1</u>	Tuberous sclerosis Bourneville's disease Epiloia	759.5
<u>Q85.8</u>	Other phakomatoses, NEC (must specify) Peutz-Jeghers Syndrome Sturge-Weber (-Dimitri) syndrome Von Hippel-Lindau syndrome	759.6

	Neuro-cutaneous syndrome Excludes 1: Meckel-Gruber syndrome (Q61.9)	
Q85.9	Phakomatosis, unspecified Hamartosis, NOS Hamartoma of retina Vascular hamartoma of skin	759.6
Q86	Congenital malformations syndromes due to known exogenous causes, NEC Excludes 2: iodine-deficiency-related hypothyroidism (E00-E02), nonteratogenic effects of substances transmitted via placenta or breast milk (P04.-)	
Q86.0	Fetal alcohol syndrome (dysmorphic) (reportable to age 10)	760.71
Q86.1	Fetal hydantoin syndrome Meadow's syndrome	760.77
Q86.2	Dysmorphism due to warfarin	760.79
Q86.8	Other congenital malformation syndromes due to known exogenous causes	760.76
Q87	Other specified congenital malformation syndromes affecting multiple systems **Use Additional code(s) to identify all associated manifestations	
Q87.0	Congenital malformation syndromes predominantly affecting facial appearance (must specify) Acrocephalopolysyndactyly Acrocephalosyndactyly [Apert syndrome] Branchio oto renal syndrome [Melnick-Fraser syndrome] Carpenter's syndrome Cryptophthalmos syndrome Cyclopia Freeman Sheldon syndrome Goldenhar syndrome (oculo-auriculovertebral) Gorlin-Chaudhry-Moss syndrome Hallerman-Streiff syndrome Moebius syndrome Oro-facial-digital syndrome Oto palato digital (OPD) syndrome Pfeiffer syndrome Pierre Robin syndrome Saethre Chotzen syndrome Whistling face Antley-Bixler Branchio-oculo-facial syndrome	755.55 756.0 759.89
Q87.1	Congenital malformation syndromes predominantly associated w/ short stature (must specify)	
Q87.11	Prader-Willi syndrome	759.81
Q87.19	Other congenital malformation syndromes predominantly associated w/ short stature (must specify) Aarskog syndrome Bonnevie-Ullrich syndrome Cornelia de Lange syndrome Cockayne syndrome De Lange syndrome Dubowitz syndrome Noonan syndrome Robinow-Silverman-Smith syndrome Russell-Silver syndrome Seckel syndrome Sjogren-Larsson syndrome	759.89
Q87.2	Congenital malformation syndromes predominantly involving limbs (must specify) Holt-Oram syndrome Klippel-Trenaunay-Weber syndrome Nail patella syndrome Rubinstein-Taybi syndrome	759.89

	Sirenomelia syndrome Thrombocytopenia with absent radius [TAR] syndrome VATER syndrome	
Q87.3	Congenital malformation syndromes involving early overgrowth (must specify) Beckwith-Wiedemann syndrome Proteus syndrome Sotos syndrome Weaver syndrome	759.89
Q87.4	Marfan's syndrome	
Q87.40	Marfan's syndrome, unspecified	759.82
Q87.41	Marfan's syndrome with cardiovascular manifestations	
Q87.410	Marfan's syndrome with aortic dilation	759.82
Q87.418	Marfan's syndrome with other cardiovascular manifestations	759.82
Q87.42	Marfan's syndrome with ocular manifestations	759.82
Q87.43	Marfan's syndrome with skeletal manifestation	759.82
Q87.5	Other congenital malformation syndromes with other skeletal changes	759.89
Q87.8	Other specified congenital malformation syndromes, NEC Excludes 1: Zellweger syndrome (E71.510)	
Q87.81	Alport syndrome Hereditary nephritis	759.89
Q87.82	Arterial tortuosity syndrome	759.89
Q87.89	Other specified congenital malformation syndromes, NEC (must specify) Basal cell nevus syndrome Gorlin syndrome Laurence-Moon (-Bardet)-Biedl syndrome	759.89
Q89	Other congenital malformations, NEC	
Q89.0	Congenital absence and malformations of spleen Excludes 1: isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6)	
Q89.01	Asplenia (congenital)	759.0
Q89.09	Congenital malformations of spleen Congenital splenomegaly	759.0
Q89.1	Congenital malformations of adrenal gland Excludes 1: adrenogenital disorders (E25.-), congenital adrenal hyperplasia (E25.0)	759.1
Q89.2	Congenital malformations of other endocrine glands Congenital malformation of parathyroid or thyroid gland Persistent thyroglossal duct Thyroglossal cyst Excludes 1: congenital goiter (E03.0), congenital hypothyroidism (E03.1)	759.2
Q89.3	Situs inversus Dextrocardia with situs inversus Mirror-image atrial arrangement with situs inversus Situs inversus or transversus abdominalis Situs inversus or transversus thoracis Transposition of abdominal viscera Transposition of thoracic viscera Excludes 1: dextrocardia NOS (Q24.0)	759.3
Q89.4	Conjoined twins Craniopagus Cicephaly Pygopagus Thoracopagus	759.4
Q89.7	Multiple congenital malformations, NEC (try not to code) Multiple congenital anomalies, NOS Multiple congenital deformities, NOS Excludes 1: congenital malformation syndromes affecting multiple systems (Q87.-)	759.7
Q89.8	Other specified congenital malformations (must specify) Charge syndrome	759.89

	Kabuki syndrome Stickler syndrome Williams syndrome Use additional code(s) to identify all associated manifestations	
Q89.9	Congenital malformation, unspecified (try not to code; must specify)	759.9

Chromosomal Abnormalities, Not Elsewhere Classified (NEC) (Q90-Q99)

**** Please report karyotype (chromosome test results) in addition to the ICD code!**

ICD-10	Description	ICD-9
Q90	Trisomy 21, Down Syndrome	
Q90.0	Trisomy 21, nonmosaicism (meiotic nondisjunction) Karyotype trisomy 21 (47,XX or XY,+21)	758.0
Q90.1	Trisomy 21, mosaicism (mitotic nondisjunction) (46,XX/47,XX+21 or 46,XY/47,XY+21)	758.0
Q90.2	Trisomy 21, translocation t(13:21), t(14:21), t(15:21), t(21:22) Translocation trisomy duplication of a 21[t(21:21)]	758.0
Q90.9	Down syndrome, unspecified Trisomy 21, NOS	758.0
Q91	Trisomy 18 and Trisomy 13	
Q91.0	Trisomy 18, nonmosaicism (meiotic nondisjunction) Edwards syndrome, karyotype trisomy 18(47,XX or XY+18)	758.2
Q91.1	Trisomy 18, mosaicism (mitotic nondisjunction) (46,XX/47,XX+18 or 46,XY/47,XY+18)	758.2
Q91.2	Trisomy 18, translocation	758.2
Q91.3	Trisomy 18, unspecified Trisomy 18, complete Edwards syndrome, NOS	758.2
Q91.4	Trisomy 13, nonmosaicism (meiotic nondisjunction) Patau syndrome, karyotype trisomy 13(47,XX or XY,+13)	758.1
Q91.5	Trisomy 13, mosaicism nondisjunction (46,XX/47, XX+13 or 46,XY/47,XY+13)	758.1
Q91.6	Trisomy 13, (translocation) t(13:14), t(13:21), t(13:15), t(13:22), t(13:13)	758.1
Q91.7	Trisomy 13, unspecified Patau syndrome, NOS	758.1
Q92	Other trisomies and partial trisomies of the autosome, NEC Includes: unbalanced translocations and insertions Excludes 1: trisomies of chromosomes 13, 18, 21 (Q90-Q91)	
Q92.0	Whole chromosomes trisomy, nonmosaicism (meiotic nondisjunction) <ul style="list-style-type: none"> • Trisomy 1 • Trisomy 2 • Trisomy 3 • Trisomy 4 • Trisomy 5 • Trisomy 6 • Trisomy 7 • Trisomy 8 • Trisomy 9 • Trisomy 10 • Trisomy 11 • Trisomy 12 • Trisomy 14 • Trisomy 15 • Trisomy 16 • Trisomy 17 • Trisomy 19 • Trisomy 20 • Trisomy 22 	758.5
Q92.1	Whole chromosome trisomy, mosaicism (mitotic nondisjunction) Mosaic trisomy of any chromosome except 21, 13 or 18	758.5
Q92.2	Partial trisomy Less than whole arm duplicated Whole arm or more duplicated Excludes 1: partial trisomy due to unbalanced translocation (Q92.5)	758.2
Q92.5	Duplications with other complex rearrangements Partial trisomy due to unbalanced translocations Partial trisomy syndromes (unbalanced, not defined) **Also code any associated deletions due to unbalanced translocations, inversions and insertions	758.5
Q92.6	Marker chromosomes Trisomies due to dicentrics	

	Trisomies due to extra rings Trisomies due to isochromosomes Individual with marker heterochromatin	
Q92.61	Marker chromosomes in normal individual	758.5
Q92.62	Marker chromosomes in abnormal individual	758.5
Q92.7	Triploidy and polyploidy <i>Triploidy, 69 chromosomes</i> <i>Other Polyploidy (96 chromosomes or other multiples of 23, except 46,69)</i>	758.5
Q92.8	Other specified trisomies and partial trisomies of autosomes (chromosomes 1-22) (must specify) Duplication identified by fluorescence in situ hybridization (FISH) Duplications identified by in situ hybridization (ISH) Duplications seen only at prometaphase	758.5
Q92.9	Trisomy and partial trisomy of autosomes, unspecified <i>Trisomy, NOS</i>	758.5
Q93	Monosomies and deletions from the autosomes, NEC	
Q93.0	Whole chromosomes monosomy, nonmosaicism (meiotic nondisjunction) <i>E.g. (45, XX, -21) or (45,XY,-21) for any chromosome #1-22</i>	758.5
Q93.1	Whole chromosome monosomy, mosaicism (mitotic nondisjunction) <i>Monosomy 21 mosaicism: e.g.: (46,XX/45,XX,-21) or (46,XY/45,XY,-21)</i> <i>Monosomy of other whole chromosomes #1-22 in mosaic form</i>	758.5
Q93.2	Chromosome replaced with ring, dicentric or isochromosomes	758.5
Q93.3	Deletion of short arm of chromosome 4 4p deletion syndrome Del4p15 Deletion 4p15-16 Wolff-Hirschorn syndrome	758.39
Q93.4	Deletion of short arm of chromosome 5 Cri-du-chat syndrome (5p-)	758.31
Q93.5	Other deletions of part of a chromosome	
Q93.51	Angelman syndrome	
Q93.59	Other deletions of part of a chromosome <i>Antimongolism syndrome: partial or total deletion of 21 [del (21)]</i> <i>Deletion of long arm 13: (del(13q)) or 13q-</i> <i>Deletion of long arm 17: (del(17q)) or 17q-</i> <i>Deletion of long arm 18: (del(18q)) or 18q-</i> <i>Deletion of short arm 17: (del(17p)) or 17p-</i> <i>Deletion of short arm 18: (del(18p)) or 18p-</i>	
Q93.7	Deletions with other complex rearrangements Deletions due to unbalanced translocations, inversions and insertions <i>**Code also any associated duplications due to unbalanced translocations, inversions and insertions</i>	758.39
Q93.8	Other deletions from the autosomes	
Q93.81	Velo-cardio-facial syndrome Deletion 22q11.2	758.32
Q93.82	Williams syndrome	
Q93.88	Other microdeletions (must specify) Miller-Dieker syndrome Smith-Magenis syndrome	758.33
Q93.89	Other deletions from the autosomes (chromosomes 1-22) (must specify) Deletions identified by fluorescence in situ hybridization (FISH) Deletions identified by in situ hybridization (IS) Deletions seen only at prometaphase	758.39
Q93.9	Deletion from autosomes, unspecified (try not to use) Autosomal deletion syndrome <i>Chromosomal deletion</i>	758.39

Q95	Balanced rearrangements and structural markers, NEC Includes: Robertsonian and balanced reciprocal translocations and insertions	
Q95.0	Balanced translocation and insertion in normal individual	758.4
Q95.1	Chromosome inversion in normal individual	758.4
Q95.2	Balanced autosomal rearrangement in abnormal individual	758.5
Q95.3	Balanced sex/autosomal rearrangement in abnormal individual	758.5
Q95.5	Individual with autosomal fragile site <i>Autosomal fragile site [fra (_)]</i>	758.4
Q95.8	Other balanced rearrangements and structural markers (must specify)	758.4
Q95.9	Balanced rearrangement and structural marker, unspecified	758.4
Q96	Turner's syndrome XO syndrome Bonnevie-Ullrich syndrome [Only applicable to female patients.] Excludes 1: Noonan syndrome (Q87.19), pure gonadal dysgenesis (Q99.1)	
Q96.0	Karyotype 45,X <i>Turner's phenotype, karyotype 45,X</i>	758.6
Q96.1	Karyotype 46, X iso (Xq) Karyotype 46, isochromosome Xq	758.6
Q96.2	Karyotype 46, X with abnormal sex chromosome, except iso (Xq) Karyotype 46, X with abnormal sex chromosome, except isochromosome Xq <i>Turner syndrome variant, [46,X,r(X)]</i> <i>Turner syndrome variant, [46,Xdel(Xq)]</i> <i>Turner syndrome variant, [46,X,del(Xp)]</i> [Only applicable to female patients.]	758.6
Q96.3	Mosaicism, 45, X/46, XX or XY [Only applicable to female patients.]	758.6
Q96.4	Mosaicism, 45, X/other cell line(s) with abnormal sex chromosome [Only applicable to female patients.]	756.6
Q96.8	Other variants of Turner's syndrome	758.6
Q96.9	Turner's syndrome, unspecified Turner Syndrome, karyotype unspecified	758.6
Q97	Other sex chromosome abnormalities, female phenotype, NEC [Only applicable to female patients.] Excludes 1: Turner's syndrome (Q96.-)	
Q97.0	Karyotype 47, XXX	758.81
Q97.1	Female with more than three X chromosomes	758.81
Q97.2	Mosaicism, lines with various numbers of X chromosomes	758.81
Q97.3	Female with 46, XY karyotype	758.81
Q97.8	Other specified sex chromosome abnormalities, female phenotype (must specify)	758.81
Q97.9	Sex chromosome abnormalities, female phenotype unspecified	758.81
Q98	Other sex chromosome abnormalities, male phenotype, NEC [Only applicable to male patients.]	
Q98.0	Klinefelter syndrome karyotype 47, XXY	758.7
Q98.1	Klinefelter syndrome, male with more than two X chromosome <i>48,XXX</i> <i>48,XXXX</i> <i>49,XXXXY</i>	758.7
Q98.3	Other male with 46, XX karyotype	758.7
Q98.4	Klinefelter syndrome, unspecified	758.7
Q98.5	Karyotype 47, XYY	758.81
Q98.6	Male with structurally abnormal sex chromosome	758.81
Q98.7	Male with sex chromosome mosaicism <i>Klinefelter syndrome mosaic (46,XY/47,XXY)</i> <i>Mosaic X/XY, 45X/46XY</i> <i>Mosaic XY/XXY, 46XY/47XXY</i> <i>Mosaic including XXXXY,49XXXXY</i>	758.81

	Mosaic XYY male[46,XY/47XYY] Sex chromosomes mosaicism	
<u>Q98.8</u>	Other specified sex chromosome abnormalities, male phenotype (must specify)	758.81
<u>Q98.9</u>	Sex chromosome abnormality, male phenotype unspecified	758.81
<u>Q99</u>	Other chromosome abnormalities, NEC	
<u>Q99.0</u>	Chimera 46, XX/46, XY Chimera 46, XX/46, XY true hermaphrodite	758.81
<u>Q99.1</u>	46, XX true hermaphrodite 46, XX with streak gonads 46, XY with streak gonads Pure gonadal dysgenesis	758.81
<u>Q99.2</u>	Fragile X chromosome Fragile X syndrome	759.83
<u>Q99.8</u>	Other specified chromosome abnormalities (must specify) Accessory autosomes, NOS Additional chromosome(s), NOS Deletion of chromosome(s), NOS Derivative chromosomes, NOS [not X or Y;der (_)] Double minute chromosomes (+dmidn) Duplication of chromosome(s), NOS Insertion, autosome [inv (_)] Inversion, autosome [inv (_)] Isochromosome, autosome [I (_)] Mosaicism, NOS Other specified anomaly of autosomes, includes satellites, stalks, excess Unspecified sex chromosome anomaly Variable region, autosome [var (_)]	758.89
<u>Q99.9</u>	Chromosomal abnormality, unspecified (try not to use)	758.9

Other Selected Reportable Conditions

ICD-10	Notes & Description	ICD-9
A50	Congenital syphilis	
A50.0	Early congenital syphilis, symptomatic	
A50.01	Early congenital syphilitic oculopathy	
A50.02	Early congenital syphilitic osteochondropathy	
A50.03	Early congenital syphilitic pharyngitis	
A50.04	Early congenital syphilitic pneumonia	
A50.05	Early congenital syphilitic rhinitis	
A50.06	Early cutaneous congenital syphilis	
A50.07	Early mucocutaneous congenital syphilis	
A50.08	Early visceral congenital syphilis	
A50.09	Other early congenital syphilis, symptomatic	
A50.1	Early congenital syphilis, latent	
A50.2	Early congenital syphilis, unspecified	
A50.3	Late congenital syphilitic oculopathy	
A50.30	Late congenital syphilitic oculopathy, unspecified	
A50.31	Late congenital syphilitic interstitial keratitis	
A50.32	Late congenital syphilitic chorioretinitis	
A50.39	Other late congenital syphilitic oculopathy	
A50.4	Late congenital neurosyphilis [juvenile neurosyphilis]	
A50.40	Late congenital neurosyphilis, unspecified	
A50.41	Late congenital syphilitic meningitis	
A50.42	Late congenital syphilitic encephalitis	
A50.43	Late congenital syphilitic polyneuropathy	
A50.44	Late congenital syphilitic optic nerve atrophy	
A50.45	Juvenile general paresis Dementia paralytica juvenilis	
A50.49	Other late congenital neurosyphilis Juvenile tabes dorsalis	
A50.5	Other late congenital syphilis, symptomatic	
A50.51	Clutton's joints	
A50.52	Hutchinson's teeth	
A50.53	Hutchinson's triad	
A50.54	Late congenital cardiovascular syphilis	
A50.55	Late congenital syphilitic arthropathy	
A50.56	Late congenital syphilitic osteochondropathy	
A50.57	Syphilitic saddle nose	
A50.59	Other late congenital syphilis, symptomatic	
A50.6	Late congenital syphilis, latent	
A50.7	Late congenital syphilis, unspecified	
A50.9	Congenital syphilis, unspecified	
D82	Immunodeficiency associated with other major defects Excludes 1: ataxia telangiectasia [Louis-Bar] (G11.3)	
D82.1	Di George's syndrome Also code 22q deletion separately if reported Pharyngeal pouch syndrome Thymic alymphoplasia Thymic aplasia or hypoplasia with immunodeficiency	279.11
E25	Adrenogenital disorders Please report cytogenetic diagnosis Includes: adrenogenital syndromes, virilizing or feminizing, whether acquired or due to adrenal hyperplasia consequent on inborn enzyme defects in hormone synthesis, Female adrenal pseudohermaphroditism, Female heterosexual precocious pseudopuberty, Male isosexual precocious pseudopuberty, Male macrogenitosomia praecox, Male sexual precocity with adrenal hyperplasia, Male virilization (female)	

	Excludes 1: indeterminate sex and pseudohermaphroditism (Q56), chromosomal abnormalities (Q90-Q99)	
E25.0	Congenital adrenogenital disorders associated with enzyme deficiency Congenital adrenal hyperplasia (CAH) 21-Hydroxylase deficiency Salt-losing congenital adrenal hyperplasia	255.2
E25.8	Other adrenogenital disorders idiopathic adrenogenital disorder	255.2
E25.9	Adrenogenital disorder, unspecified Adrenogenital syndrome NOS	255.2
E34	Other endocrine disorders ** only report if congenital or genetic Excludes 1: pseudohypoparathyroidism (E20.1)	
E34.3	Short stature due to endocrine disorder Laron-type short stature Excludes 1: achondroplastic short stature (Q77.4), hypochondroplastic short stature (Q77.4), nutritional short stature (E45), pituitary short stature (E23.0), progeria (E34.8), renal short stature (N25.0), Russell-Silver syndrome (Q87.19), short-limbed stature with immunodeficiency (D82.2), short stature in specific dysmorphic syndromes - code to syndrome, short stature, NOS (R62.52)	259.4
E34.5	Androgen insensitivity syndrome	
E34.50	Androgen insensitivity syndrome, unspecified	259.5
E34.51	Complete androgen insensitivity syndrome Complete androgen insensitivity de Quervain syndrome Goldberg-Maxwell syndrome Testicular feminization	259.5
E34.52	Partial androgen insensitivity syndrome Partial androgen insensitivity Reifenstein syndrome	259.5
E78	Disorders of lipoprotein metabolism and other lipidemias Excludes 1: sphingolipidosis (E75.0-E75.3)	
E78.72	Smith-Lemli-Optiz syndrome	759.89
G12	Spinal muscular atrophy and related syndromes	
G12.0	Infantile spinal muscular atrophy, type I Werdnig-Hoffman disease	335.0
G12.1	Other inherited spinal muscular atrophy (must specify) Adult form spinal muscular atrophy Childhood form, type II spinal muscular atrophy Distal spinal muscular atrophy Juvenile form, type III spinal muscular atrophy [Kugelberg-Welander] Progressive bulbar palsy of childhood [Fazio-Londe] Scapuloperoneal form spinal muscular atrophy	335.11
G12.9	Spinal muscular atrophy, unspecified	335.10
G60	Hereditary and idiopathic neuropathy	
G60.0	Hereditary motor and sensory neuropathy Charcot-Marie-Tooth disease Déjérine-Sottas disease Hereditary motor and sensory neuropathy, types I-IV Hypertrophic neuropathy of infancy Peroneal muscular atrophy (axonal type) (hypertrophic type) Roussy-Levy syndrome	356.0
G60.1	Refsum disease Infantile Refsum disease	356.3
G60.2	Neuropathy in association with hereditary ataxia	356.0
G71	Primary disorders of muscles Excludes 2: arthrogryposis multiplex congenita (Q74.3), metabolic disorders (E70-E88), myositis (M60.-)	

G71.0	Muscular dystrophy	
G71.00	Muscular dystrophy, unspecified (must specify) (reportable to age 10)	359.1
G71.01	Duchenne or Becker muscular dystrophy (reportable to age 10) Autosomal recessive, childhood type, muscular dystrophy resembling Duchenne or Becker muscular dystrophy Benign [Becker] muscular dystrophy Severe [Duchenne] muscular dystrophy	359.1
G71.02	Facioscapulohumeral muscular dystrophy (reportable to age 10) Scapulohumeral muscular dystrophy	359.1
G71.09	Other specified muscular dystrophies (must specify) (reportable to age 10) Benign scapuloperoneal muscular dystrophy with early contractures [Emery-Dreifuss] Congenital muscular dystrophy NOS Congenital muscular dystrophy with specific morphological abnormalities of the muscle fiber Distal muscular dystrophy Limb-girdle muscular dystrophy Ocular muscular dystrophy Oculopharyngeal muscular dystrophy Scapuloperoneal muscular dystrophy	359.1
G71.1	Myotonic disorders	
G71.11	Myotonic muscular dystrophy (reportable to age 10) Dystrophia myotonica [Steinert] Myotonia atrophica Myotonic dystrophy Proximal myotonic myopathy (PROMM) Steinert disease	359.21
G71.12	Myotonia congenita Acetazolamide responsive myotonia congenita Dominant myotonia congenita [Thomsen disease] Myotonia levior Recessive myotonia congenita [Becker disease]	359.22
G71.13	Myotonic chondrodystrophy Chondrodystrophic myotonia Congenital myotonic chondrodystrophy Schwartz-Jampel disease	359.23
G71.19	Other specified myotonic disorders (must specify) Isaac-Mertens syndrome Myotonia fluctuans Myotonia permanens Neuromyotonia [Isaacs] Paramyotonia congenita (of von Eulenburg) Pseudomyotonia Symptomatic myotonia	359.29
G71.2	Congenital myopathies Excludes 1: arthrogryposis multiplex congenita (Q74.3)	359.0
G71.20	Congenital myopathy, unspecified Fiber-type disproportion	
G71.21	Nemaline myopathy	
G71.22	Centronuclear myopathy	
G71.220	X-linked myotubular myopathy Myotubular (centronuclear) myopathy	
G71.228	Other centronuclear myopathy Autosomal centronuclear myopathy Autosomal dominant centronuclear myopathy Autosomal recessive centronuclear myopathy Centronuclear myopathy, NOS	
G71.29	Other congenital myopathy (must specify)	

	Central core disease Minicore disease Multicore disease Multiminicore disease	
G71.3	Mitochondrial myopathy, not elsewhere classified Excludes 1: Kearns-Sayre syndrome (H49.81), Leber's disease (H47.21), Leigh's encephalopathy (G31.82), mitochondrial metabolism disorders (E88.4.-), Reye's syndrome (G93.7)	359.89
G71.8	Other primary disorders of muscles Congenital hemihypertrophy	359.89
G71.9	Primary disorder of muscle, unspecified Hereditary myopathy, NOS	359.9
G80	Cerebral Palsy is Not Reportable	
K40	Inguinal Hernia (report for female only) Includes: bubonocele, direct inguinal hernia, double inguinal hernia, indirect inguinal hernia, inguinal hernia NOS, oblique inguinal hernia, scrotal hernia	
K40.00	Bilateral inguinal hernia, with obstruction, without gangrene, not specified as recurrent Bilateral inguinal hernia, with obstruction, without gangrene, NOS	550.12
K40.1	Bilateral inguinal hernia, with gangrene (report for female only)	
K40.10	Bilateral inguinal hernia, with gangrene, not specified as recurrent Bilateral inguinal hernia, with gangrene, NOS	550.02
K40.2	Bilateral inguinal hernia, without obstruction or gangrene (female only)	
K40.20	Bilateral inguinal hernia, without obstruction or gangrene, not specified as recurrent Bilateral inguinal hernia, NOS	550.92
K40.3	Unilateral inguinal hernia, with obstruction, without gangrene (female only) <ul style="list-style-type: none"> • Inguinal hernia (unilateral) causing obstruction without gangrene • Incarcerated inguinal hernia (unilateral) without gangrene • Irreducible inguinal hernia (unilateral) without gangrene • Strangulated inguinal hernia (unilateral) without gangrene Indicate right, left, or unspecified	
K40.30	Unilateral inguinal hernia, with obstruction, without gangrene, not specified as recurrent Inguinal hernia, with obstruction, NOS Unilateral inguinal hernia, with obstruction, without gangrene, NOS	550.1
K40.40	Unilateral inguinal hernia, with gangrene, not specified as recurrent Inguinal hernia with gangrene, NOS Unilateral inguinal hernia with gangrene, NOS	550
K40.9	Unilateral inguinal hernia, without obstruction or gangrene (female only) Indicate right, left, or unspecified	
K40.90	Unilateral inguinal hernia, without obstruction or gangrene, not specified as recurrent Direct inguinal hernia Indirect inguinal hernia Inguinal hernia, NOS Left direct inguinal hernia Left indirect inguinal hernia Left inguinal hernia Right direct inguinal hernia Right indirect inguinal hernia Right inguinal hernia	550.9
K41	Femoral hernia (female only)	
K41.0	Bilateral femoral hernia, with obstruction, without gangrene (female only) <ul style="list-style-type: none"> • Femoral hernia (bilateral) causing obstruction, without gangrene • Incarcerated femoral hernia (bilateral), without gangrene • Irreducible femoral hernia (bilateral), without gangrene • Strangulated femoral hernia (bilateral), without gangrene 	
K41.00	Bilateral femoral hernia, with obstruction, without gangrene, not specified as recurrent Bilateral femoral hernia, with obstruction, without gangrene, NOS	552.02
K41.1	Bilateral femoral hernia, with gangrene (female only)	

<u>K41.10</u>	Bilateral femoral hernia, with gangrene, not specified as recurrent Bilateral femoral hernia, with gangrene, NOS	551.02
<u>K41.2</u>	Bilateral femoral hernia, without obstruction or gangrene (female only)	
<u>K41.20</u>	Bilateral femoral hernia, without obstruction or gangrene, not specified as recurrent Bilateral femoral hernia, NOS	553.02
<u>K41.3</u>	Unilateral femoral hernia, with obstruction, without gangrene (female only) <ul style="list-style-type: none"> Femoral hernia (unilateral) causing obstruction, without gangrene Incarcerated femoral hernia (unilateral), without gangrene Irreducible femoral hernia (unilateral), without gangrene Strangulated femoral hernia (unilateral), without gangrene Indicate right, left, or unspecified	
<u>K41.30</u>	Unilateral femoral hernia, with obstruction, without gangrene, not specified as recurrent Femoral hernia, with obstruction NOS Unilateral femoral hernia, with obstruction NOS	552
<u>K41.4</u>	Unilateral femoral hernia, with gangrene (female only) Indicate right, left, or unspecified	
<u>K41.40</u>	Unilateral femoral hernia, with gangrene, not specified as recurrent Femoral hernia, with gangrene NOS Unilateral femoral hernia, with gangrene NOS	551
<u>K41.9</u>	Unilateral femoral hernia, without obstruction or gangrene (female only) Indicate right, left, or unspecified	
<u>K41.90</u>	Unilateral femoral hernia, without obstruction or gangrene, not specified as recurrent Femoral hernia, NOS Unilateral femoral hernia, NOS	553
<u>L05</u>	(m) Pilonidal cyst and sinus	
<u>L05.9</u>	(m) Pilonidal cyst and sinus without abscess	
<u>L05.91</u>	(m) Pilonidal cyst without abscess (m) Pilonidal dimple (m) Pilonidal cyst NOS (m) Postanal dimple Excludes 2: congenital sacral dimple (Q82.6), parasacral dimple (Q82.6)	685.1
<u>L05.92</u>	(m) Pilonidal sinus without abscess (m) Coccygeal fistula (m) Coccygeal sinus without abscess (m) Pilonidal fistula	685.1
<u>L81</u>	(m) Other disorders of pigmentation Excludes 1: birthmark NOS (Q82.5), Peutz-Jeghers syndrome (Q85.8) Excludes 2: nevus - see Alphabetical Index	
<u>L81.3</u>	(m) Café au lait spots	709.09
<u>L81.4</u>	(m) Other melanin hyperpigmentation	709.09
<u>L81.9</u>	(m) Disorder of pigmentation, unspecified	709
<u>M26</u>	(m) Dentofacial anomalies [including malocclusion]	
<u>M26.0</u>	(m) Major anomalies of jaw size Excludes 1: acromegaly (E22.0), Robin's syndrome (Q87.0)	
<u>M26.01</u>	(m) Maxillary hyperplasia (m) Prominent maxilla	524.01
<u>M26.02</u>	(m) Maxillary hypoplasia	524.03
<u>M26.03</u>	(m) Mandibular hyperplasia	524.02
<u>M26.04</u>	(m) Mandibular hypoplasia	524.04
<u>M26.05</u>	(m) Macrogenia	524.05
<u>M26.06</u>	(m) Microgenia	524.06
<u>M26.09</u>	Micrognathia Macrognathia (m) Other specified anomalies of jaw (specify)	524.09
<u>M26.1</u>	Anomalies of jaw-cranial base relationship	
<u>M26.19</u>	Other specified anomalies of jaw-cranial base relationship (must specify)	524.19

	Retrognathia (m) Congenital prognathism (m) Mandibular prognathism (m) Maxillary retrognathia (m) Maxillary retrognathism	
P02	Newborn (suspected to be) affected by complications of placenta, cord and membranes Includes: conditions that have their origin in the fetal or perinatal period (before birth through the first 28 days after birth) even if morbidity occurs later Code first any current condition in newborn	
P02.8	Newborn (suspected to be) affected by other abnormalities of membranes Amniotic band syndrome Amniotic bands (constricting bands) Streeter's (Simonart's) bands P02.8 is only applicable to newborns of age 0 years.	762.8
P35	Congenital viral diseases Includes: infections acquired in utero or during birth Only applicable to newborns of age 0 years.	
P35.0	Congenital Rubella Syndrome Rubella, congenital Congenital rubella pneumonitis	771.0
P35.1	Congenital cytomegalovirus infection (CMV) Cytomegalovirus infection, congenital	771.1
P35.2	Congenital herpesviral [herpes simplex] infection Herpes simplex Encephalitis herpes Meningoencephalitis herpes	771.2
P35.3	Congenital viral hepatitis	771.2
P35.4	Congenital Zika virus disease Use Additional Code to identify manifestations of congenital Zika virus disease	
P35.8	Congenital infection, other specified Congenital varicella [chickenpox]	771.2
P35.9	Congenital viral disease, unspecified	771.2
P37	Other congenital infectious and parasitic diseases Excludes 2: congenital syphilis (A50.), infectious neonatal diarrhea (A00-A09), necrotizing enterocolitis in newborn (P77.-), noninfectious neonatal diarrhea (P78.3), ophthalmia neonatorum due to gonococcus (A54.31), tetanus neonatorum (A33)	
P37.0	Congenital tuberculosis P37.0 is only applicable to newborns of age 0 years.	771.2
P37.1	Congenital toxoplasmosis Hydrocephalus due to congenital toxoplasmosis P37.1 is only applicable to newborns of age 0 years.	771.2
P96	Other conditions originating in the perinatal period	
P96.1	Neonatal withdrawal symptoms from maternal use of drugs of addiction	

Stillbirth Codes (O36.4, Z37)

ICD-10	Notes & Description	ICD-9
<u>O36.4</u>	Maternal care for intrauterine death <ul style="list-style-type: none"> • Maternal care for intrauterine fetal death NOS • Maternal care for intrauterine fetal death after completion of 20 weeks of gestation • Maternal care for late fetal death • Maternal care for missed delivery <p>Only applicable to female maternity patients aged 12 - 55 years inclusive. Should only be used on the maternal record - not on the newborn record. Use a code under <u>Z3A</u> to document the exact week during the pregnancy.</p> <p>Excludes 1: missed abortion (<u>O02.1</u>), stillbirth (<u>P95</u>)</p>	
<u>O36.4XX0</u>	Maternal care for intrauterine death, not applicable or unspecified Dead fetus in utero Fetal death from asphyxia and/or anoxia, not clear if noted before or after onset of labor Fetal death from asphyxia or anoxia Pregnancy loss	656.40 656.41 656.43
<u>O36.4XX1</u>	Maternal care for intrauterine death, fetus 1	656.41 656.43
<u>O36.4XX2</u>	Maternal care for intrauterine death, fetus 2	656.41 656.43
<u>O36.4XX3</u>	Maternal care for intrauterine death, fetus 3	656.41 656.43
<u>O36.4XX4</u>	Maternal care for intrauterine death, fetus 4	656.41 656.43
<u>O36.4XX5</u>	Maternal care for intrauterine death, fetus 5	656.41 656.43
<u>O36.4XX9</u>	Maternal care for intrauterine death, other fetus	656.41 656.43
<u>P95</u>	Stillbirth Fetal death of unspecified cause Excludes 1: maternal care for intrauterine death (<u>O36.4</u>), missed abortion (<u>O02.1</u>), outcome of delivery, stillbirth (<u>Z37.1</u> , <u>Z37.3</u> , <u>Z37.4</u> , <u>Z37.7</u>)	779.6
<u>Z37.1</u>	Outcome of delivery, single stillborn	V27.1
<u>Z37.3</u>	Outcome of delivery, twins, one liveborn and one stillborn	V27.3
<u>Z37.4</u>	Outcome of delivery, twins, both stillborn	V27.4
<u>Z37.6</u>	Outcome of delivery, other multiple birth, some liveborn	V27.6
<u>Z37.60</u>	Multiple births, unspecified, some liveborn	V27.6
<u>Z37.61</u>	Triplets, some liveborn	V27.6
<u>Z37.62</u>	Quadruplets, some liveborn	V27.6
<u>Z37.63</u>	Quintuplets, some liveborn	V27.6
<u>Z37.64</u>	Sextuplets, some liveborn	V27.6
<u>Z37.69</u>	Other multiple births, some liveborn	V27.6
<u>Z37.7</u>	Outcome of delivery, other multiple birth, all stillborn	V27.7

Revision History

Since the December 2015 version, we added “(must specify)” next to some of the non-specific narrative descriptions. In addition, we added more synonyms to the list of narrative descriptions.

Added/modified Codes due to October 2020 ICD10 revision:

A50	A50.2	A50.45	A50.7	G71.02
A50.0	A50.3	A50.49	A50.9	G71.09
A50.01	A50.30	A50.5	Q35-Q37	G71.11
A50.02	A50.31	A50.51	Q51.20	G71.2
A50.03	A50.32	A50.52	Q51.21	G71.20
A50.04	A50.39	A50.53	Q51.22	G71.21
A50.05	A50.4	A50.54	Q51.28	G71.22
A50.06	A50.40	A50.55	Q82.8	G71.220
A50.07	A50.41	A50.56	Q86.0	G71.228
A50.08	A50.42	A50.57	A50.9	G71.29
A50.09	A50.43	A50.59	G71.00	P96.1
A50.1	A50.44	A50.6	G71.01	

Added/modified Codes due to October 2019 ICD10 revision:

Q02	Q66.211	Q66.32	Q66.90	Q82.8
Q66.0	Q66.212	Q66.4	Q66.91	Q87.1
Q66.00	Q66.219	Q66.40	Q66.92	Q87.11
Q66.01	Q66.22	Q66.41	Q79.6	Q87.19
Q66.02	Q66.221	Q66.42	Q79.60	Q96
Q66.1	Q66.222	Q66.7	Q79.61	E34.3
Q66.10	Q66.229	Q66.70	Q79.62	
Q66.11	Q66.3	Q66.71	Q79.63	
Q66.12	Q66.30	Q66.72	Q79.69	
Q66.21	Q66.31	Q66.9	Q81.0	

Added/modified Codes due to October 2018 ICD10 revision:

Q02	Q51.21	Q93.51	G71.0	G71.02
Q51.2	Q51.28	Q93.59	G71.00	G71.09
Q51.20	Q93.5	Q93.82	G71.01	P35.4

Added/modified Codes due to October 2017 ICD10 revision:

Q25.49	Q53.111	Q53.13	Q53.211	Q53.23
Q53.11	Q53.112	Q53.21	Q53.212	Q82.2

Added/modified Codes due to October 2016 ICD10 revision:

Q25.2	Q25.42	Q25.48	Q52.123	Q82.6
Q25.21	Q25.43	Q25.49	Q52.124	Q87.82
Q25.29	Q25.44	Q52.12	Q52.129	
Q25.4	Q25.45	Q52.120	Q66.2	
Q25.40	Q25.46	Q52.121	Q66.21	
Q25.41	Q25.47	Q52.122	Q66.22	

Removed codes from the Other Selected Reportable Conditions section, March 2017:

- All D codes besides D82.1 Di George’s Syndrome
- E codes up until and including E23.2
- E34.8
- E70-E78.71
- E79-88.89
- G11
- G23-52.7
- H Codes
- P04

Reclassified codes (effective March 2017):

Description	Previous	Revised	Notes
Sacral dimple	L05.91	Q82.6	Description under “Excludes” for L05.91
Parasacral dimple	L05.91	Q82.6	
(m) <i>Scaphocephaly (if due to craniosynostosis)</i>	Q75.0	Q67.4	
(m) <i>Trigonocephaly, other head deformations without synostosis</i>	Q75.0	Q67.4	

Changes effective October 2018

Code	Defect	Previous classification	Current classification
Q25.6	Peripheral pulmonic stenosis (PPS)	Minor	Major, only report if $\geq 2500\text{g}$ at birth
Q55.63	Congenital torsion of penis	Major	Minor
Q55.63	Phimosis	Major	Not Reportable

- P95 moved to stillbirth section from ‘other selected reportable conditions’ section.

Additional changes effective October 2019

Code	Defect	Previous classification	Current classification
Q55.64	Hidden penis	Major	Minor
Q55.64	Buried penis	Major	Minor
Q04.6	Congenital choroid plexus cyst	Major	Minor